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Life and Change with Usher: The
experiences of diagnosis for people
with Usher syndrome.

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1 Executive summary

Usher syndrome is one of the largest causes of deafblindness in the UK but prior to this report, there was little published research on the lives of people with Usher. Sense funded a large collaborative study on Usher (Sense 2012) but this was a medical study focusing on evaluating the genetic cause of Usher in the UK. People with Usher documented their experiences in their own writings, in the news, on blogs and articles. This project was funded by Sense to be structured enquiry about the lives of people with Usher, not only when they were diagnosed but also afterwards, as people make adjustments and changes. It explores the lives of people with Usher as seen by themselves, how the effect of the changes inherent in Usher and how knowing that they have Usher has an impact on their lives, the way they lead their lives and their concepts of themselves.

It aimed to explore the questions;

- What do people with Usher think about having Usher syndrome?
- What is the effect of change on the lives of people with Usher?
- What do people with Usher remember of their diagnosis and what impact did it have on them?

1.1 Usher syndrome

Usher syndrome is a genetic condition which affects vision and hearing. There are three main types of Usher syndrome, Usher 1, 2 and 3. The traditional picture of Usher is that there are three types; 1, 2 and 3, which were primarily differentiated by the degree of deafness and the age of onset of visual problems. People with type 1 were profoundly or severely deaf from birth, had severe balance problems and the onset of visual problems was in the region of early teens. Many will have used British Sign Language (BSL) as their first language and consider themselves to be part of the cultural group of deaf people who use signs – the Deaf community (usually written with a capital D). Early cochlear implantation has changed this profile somewhat. People with Usher type

2 were moderately to severely deaf from birth, and the onset of noticeable visual problems was usually in the later teens. They did not have balance problems and they were usually users of spoken English. Usher type 3 was discovered later, and is the rarest kind in the U.K. and the hearing impairment and the visual impairment are progressive, and usually emerge in early middle age

Loss of sight is gradual and is due to a progressive eye condition called Retinitis Pigmentosa (RP). This means that vision reduces, over time, and with some degree of unpredictability. Usher is a genetic condition thus families in which there is more than one person with Usher are not uncommon.

1.1.1 Methodology

Our work was based on a case study methodology, exploring with individuals their perspective about the way in which they responded to the challenge of Usher syndrome. Qualitative data was gathered; it is not intended to be at all representative in the sense that the participants were not chosen to represent and be typical of the population. However, it allows the research to show a full picture of a range of different responses and to explore through these the implications for people with Usher syndrome. Our case studies stand alone, as each individual describes their own experience as a person with Usher but also together, as they create a multi-faceted picture of the way people with Usher conceptualise themselves and deal with life.

Interviews were the main method of gathering information. These were semi-structured, extended interviews which encouraged people to talk about their lives. Using interviews meant that we would be able to communicate with people who had visual and hearing impairments including offering equal access to people who used BSL (British Sign Language), including hands-on BSL, or SSE (Sign Supported English) or braille. Interpretation from their chosen and familiar interpreters was provided for those who wished to communicate in sign language.

We interviewed the largest cohort of people with Usher in a social research study – 44 people with Usher, 15 with Usher 1, 22 with Usher 2

and 5 with Usher 3. Of these 31 used spoken English and 11 sign (BSL, SSE or both). They came from all over the UK and were aged between 14 and 56.

We also carried out two focus group interviews; one with two people with Usher 2 (who used spoken English) and one with three participants with Usher 1 (who used BSL). We also interviewed 3 young people for a second time.

In addition, we used a standardised measure of self image; the Self Image Profile (SIP). This is a paper exercise in which people indicate on a scale from 0-6 how much they think they are like certain characteristics (happy, fit, confident) and then how much they would like to be those things, and results in a numerical score of self image and self esteem.

1.2 What is it like to have Usher in the UK today?

From our interviews and focus groups, and combined with the SIP, we present the following summary. What is it like to have Usher in the UK today? These are the themes emerging from the voices of people with Usher.

1.2.1 They enjoy life

Despite some other indications that most people with Usher might be living with high levels of depression (e.g. Miner 1995 and 1997, Bodsworth et al. 2011) we also found excitement, interest and anticipation. From the SIP it was clear that people with Ushers were not unusually pessimistic. They were doing things they wanted to do, either despite, or occasionally because of, Usher. They were successful (if sometimes worried) parents, competent employees, and good friends. They played music, socialised with others, and took part in sport. They found friends or supporters who also had Usher or joined Facebook groups. They had already accepted being deaf – becoming visually impaired was not something they wanted, but it was the hand they had been dealt. Usher was obviously part of what was going on, but it was not all of it. They did most of the things which other people in the UK did. Undoubtedly some were, or had been, depressed. Depression may have occurred when they heard about Usher, but it also happened at

moments of change, particularly those associated with mobility – such as starting to use a cane, or giving up driving.

1.2.2 They manage

They had learned to manage, and they managed. Having Usher did mean that they had to do things differently, and perhaps to give some things up. They had to learn to use specialist aids and they found ways of using technology to help them. They used public transport or guide dogs rather than having the freedom they may have had in a car. They learnt new skills, including how to tell professionals what they needed.

One of the main things they did was **turn on the lights**. Good lighting which did not create glare played a very significant part in people's ability to manage.

There were understandable and often avoidable frustrations, such as with colleagues or neighbours who did not understand what they could not, and equally important, could, do. Systems and the environment (from not getting the help they needed to overhanging branches) also frustrated them.

1.2.3 Things develop

Change is part of the natural process of life that things change. Change is not, of itself, unusual.

What was more unusual perhaps was that the changes challenged their own perceptions of themselves, that they challenged individuals' ontological security. Not knowing when things will change, and how much they will change means that they live sometimes with little continuity - a defining characteristic of this security. As they live with Usher, other things will change too, and those changes interacting with the difficulties of having Usher- moving house, having children, losing a job, make that continuity very hard. This unpredictability of life was reflected in people's accounts of 'doing things while I still can' and 'I don't know if it's worth it.'

The research also presented a cross section of time (through interviewing people of different ages) which highlighted changes in society. In this study, the especially pertinent ones are changes in the identification of the typology of Usher, changes in education and changes in diagnosis.

Gene identification has shown that more genes and a wider range of 'types' of Usher are involved than previously thought. This has challenged the symptomatic identification of Usher types 1,2,3 with the related 'timescale' of deafness, the degree of deafness, the onset of visual impairment. The genetic identification does not always match the symptomatic presentation.

Cochlear implantation and a more inclusive educational environment have led to a greater proportion of young people with Usher attending mainstream schools, and fewer of them using BSL. There is some indication that there was more recently increased openness in telling young people about Usher. This could be related to the availability of genetic testing.

1.3 The impact of diagnosis

The experience of being diagnosed with Usher was one of the key issues of the study. For some, the impact of the diagnosis was for some the start of depression and struggle. But for others it was a spur to do more, and to others it was something to put aside for the moment. .

Some people living with Usher, treated it as a thing – external to themselves, for others, as a way of being – integral to who they are. For those who treated it as a thing, Usher was something in their lives, something which they carried with them all the time, and to that extent it was part of them. However it did not define who they were, or what they did. They might have to get round it, avoid it, or sometimes face it head on and accept its presence, but they did not take it into themselves.

For others, it was a way of being. Usher was part of them, it was one of the things which identified them, which they saw as key to who they were. It was still one of the things that made them themselves. Some embraced it and would not have chosen to be different. Their life would

not be exactly the way they had envisaged it but that did not mean it had to be less.

Participants often embodied both of these positions simultaneously. Sometimes Usher was a thing – an eye condition, a hearing impairment, something which they could not do anything about, which they chose to ignore, and at other times sought to challenge. Yet at the same time Usher was central to their identity and made them who they were.

The point at which the impact of visual impairment was unavoidable was sometimes the point at which they also began to think of themselves as deaf. Their vision had changed, but so had, to them, their hearing status. For some of the Deaf people, who used BSL, at the point at which they experienced significant changes to their vision, some of them began to feel less a part of the Deaf community and less in touch with BSL. Visual impairment exacerbates hearing impairment. Mobility for a visually impaired person presents challenges, but as a dual sensory impaired person it presents far more. Communication became difficult when they could no longer lip read easily. People with Usher may not have felt deaf before, but now they were **deafblind**. Support was related to gadgets which helped hearing impaired people or visually impaired people, or strategies which helped one or the other –there was little which embraced deafblindness.

1.4 The Usherness of Usher

Throughout the interviews, three features of the lives of people with Usher syndrome were key to our understanding of how it affects people's lives. These are; Change, Predictability/Uncertainty and Diversity. In particular we drew our discussion and analysis from the themes of diagnosis, mobility, communication, education, employment, friends, family and support networks, and leisure activities.

1.4.1 Change

There were changes throughout people's lives, and for some, the changes of Usher were similar to other changes in life. But there were certain aspects of change which were crucial to the experience of people with Usher. For some, change came at the moment of diagnosis,

through changes in self perception and their sense of security. For most this happened at the point when something unavoidable changed; they stopped driving or started to read braille. These were the moments when they needed support. For some change was forced on them by the inability of the environment (e.g. school, job) to adapt to their needs. Small changes could also be significant, the need for greater lighting, or choosing to take up running hand in hand. Or they might make friends in support groups or deciding to have a baby or go quad biking **now**.

On the other hand, their communication method did not change much. All who had Usher 1, continued to use BSL after school (whether their school was oral or signing). They might be considering hands on sign. Those who were spoken English users stayed with spoken English, using hearing aids, implants or other means of amplification. While they might use signs in certain situations, they were not considering learning BSL. Some had started to use braille.

They all knew they would change – and some decided to get on with it, and cope with each change, while others hung on as long as possible in the old way before an incident forced them into something new.

The constant and unpredictable change did lead to some being depressed or frustrated. But most understood it to be part of Usher – that they had to be ready to adapt – ‘up and at ‘em’ (as Kat’s mother said).

1.4.2 Predictability/Uncertainty

Uncertainty is a theme underlying all the participants’ experiences of Usher. Predictions had been made, frequently, especially at diagnosis, but in many cases, they had turned out to be inaccurate, or at least inaccurately phrased. Some events, such as giving up driving or using a cane, were predictable, but the timing of these could not be predicted, not based on their current vision or the type of Usher with which they were diagnosed. Some decided to beat the predictions, for instance by deciding to have a family quickly, or going travelling). In this way, they challenged the predictions made and the fact that they might not be able to do things.

Uncertainty did have an effect on how people lived their lives. Interestingly, some did not want this to mean that they simply became more dependent – they did not want to become more patient or more sensitive. They wanted to take control, others looked into the future and resigned themselves to it, finding that they managed the unpredictability better if they rolled with the punches.

The behaviour of others was also unpredictable. Colleagues, family and friends had to change as well, - they thought partners might leave, or colleagues might reject them (leading to some hiding Usher from work companions) or that friends would not want to be support workers as well as friends. But in fact, in many cases, they did continue to have friends, their partners did not leave them, though colleagues in mainstream working environments did not seem, overall, to be very helpful.

1.4.3 Diversity

Finally, the participants and their experiences are and continue to be diverse. They come from diverse backgrounds and experience, they have different lifestyles, and different personal attributes. It would be more surprising if their lives were all very similar.

Participants differ in how depressed, and how isolated they are, and to what they have had to give up. They get different amounts of support. They have different experiences of clinical diagnosis. They adapt in different ways. However, there are many indications that people with Usher can do many of the things they want to – they sometimes have to find a way round it – but they can. They manage change and so take part in blind cricket, managing their own businesses, attending mother and toddler groups, undertaking adventurous sports and having successful relationships.

The progress of their visual impairment (and hearing impairment) is also very diverse, and not linked to the predictors of type or other family members' experiences. Some are managing with very little sight at as young an age as 15, while well on in their 50s, others are still using sight to get around. Individual diversity is important in managing their progressive impairment, - what was an obstacle for one person might not be for another. Levels of vision and hearing loss were not good

predictors for many of the concerns – work, school, families, activities; apart from obvious differences (such as taking part in sports for the blind rather than mainstream sports) the individual's own response was a much more important factor.

1.5 What people with Usher may need

The research brought out a number of issues which indicated what people with Usher may need, though not all wanted the same things, or wanted them at the same time. However, Usher does have an impact on school, on work, on family, and more.

At the point at which they are diagnosed, and shortly after, the stories of our participants suggested that they

- wanted to know that there are specialists. They wanted the reassurance of someone who understood Usher and who could tell them what the latest developments were.
- wanted to know what 'going blind' means. They may have been told they would 'go blind' but not understand that in fact most people with Usher do not lose all their vision very quickly, if indeed at all. Specialists cannot tell any individual what exactly will happen to them, but they can give some general advice or statistics which indicate that Usher is not necessarily what people might imagine by 'going blind'.
- wanted a level of support appropriate to their level of vision *now*. They needed information at the point of diagnosis, but they might need support later – counselling, advice, and information about resources they could use. Because Usher, unlike a medically managed illness, does not require frequent returns to doctors, they might not get in touch with anyone who can support them again for a long time, they might need to know how to get help when they did need it.

They needed support for their daily lives. Sense was a clear central provider of support for some, while others had little contact.

- Some participants did not want to be signed off as 'sick'. The experience of others with Usher suggests that with appropriate adaptations, they could continue to work. Advice on Access to

Work and employment legislation and support is probably a more useful response.

- Some participants found organisations aimed at their own age and level of visual difficulty to be their preferred support. These would include such groups as the 'Young Professionals' group from 'RP Fighting Blindness' Networks of informal support, whether meeting in person or online were as helpful for some as support from formal agencies set up to do this.
- In schools participants wanted access to support and advice from teachers specially trained in deafblindness who knew about the varied needs of people with Usher and who could tailor this to individual requirements.
- Some participants wanted detailed information on the genetic inheritance of Usher, this was particularly so for those who were planning a family.
- Participants also identified that partners, parents, friends, and siblings, were also in need of support and advice from specialist services and networks who understood the Usherness of Usher.
- Greater awareness of hidden disabilities, including communication and mobility strategies to support people with hearing and visual impairments were recommended. This was viewed as particularly important for hospital and medical staff, benefits agencies, public transport operatives, as well as the general public.
- Most significantly, overall was a desire by participants to share with other people with Usher, especially the newly diagnosed, their success, achievements, strategies for coping, and the knowledge that although it is tough, everything will be ok in the end.

2 Acknowledgements

We would like to thank a number of people for their help in preparing this report: the 44 participants with Usher syndrome who so willingly told us about their lives; the many other people who helpfully facilitated these interviews and helped in the recruitment process, in particular the participants' family members and associated professionals; the project advisory committee and colleagues at the University of Birmingham for their advice and support; Sense for funding this research.

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3 Introduction and context

3.1 Overview

3.1.1 Aims of the project

This project began when discussing what research was missing in the UK in relation to deafblind people. Usher; one the largest causes of deafblindness, was documented in medical research, with a large collaborative study (Sense 2012) but there was little research on the lives of people with Usher. They were not necessarily hidden or silent – there were news, articles, blogs about people with Usher in publications on deafblindness, and on deafness, but there was no structured enquiry about the lives of people with Usher, not only when they were diagnosed but also afterwards, as people make adjustments and changes. Sense funded this project to increase the knowledge about the lives and experiences of people with Usher.

The project was to investigate these lives and this report provides the results and discussion following this investigation. In addition, we describe the methods of the research and the methodological implications, and a literature review (non-medical) about people with Usher. At the centre are the voices of people with Usher themselves, telling their own stories.

3.2 Usher syndrome

Usher syndrome is a genetic condition which affects vision and hearing. Hearing impairment is caused by damage to the hair cells in the cochlea, or the auditory nerve, causing sensorineural deafness. In some cases both may be damaged. There are three main types of Usher syndrome, Usher 1, 2 and 3 although there are a number of subtypes delineated by genetic markers. In some types of Usher the hearing impairment is present from birth, though this may not be the case for people with Usher 3. The hearing impairment may be mild, moderate, severe, or profound.

Loss of sight is gradual and is due to a progressive eye condition called Retinitis Pigmentosa (RP). This means that vision reduces, over time, and with some degree of unpredictability. Usher is a genetic condition in which a gene or gene sequence is inherited from both parents. There is a 1 in 4 likelihood of a person having Usher if both parents are carriers of the gene, thus families in which there is more than one person with Usher are not uncommon.

The traditional picture of Usher is that there are three types; 1, 2 and 3, which were primarily differentiated by the degree of deafness and the age of onset of visual problems. People with type 1 were profoundly or severely deaf from birth, had severe balance problems and the onset of visual problems was in the region of early teens. Before cochlear implants, many will have used British Sign Language (BSL) as their first language and consider themselves to be part of the cultural group of deaf people who use signs – the Deaf community (usually written with a capital D). Early cochlear implantation has changed this profile somewhat, with more young people with Usher using spoken English. People with Usher type 2 were moderately to severely deaf from birth, and the onset of noticeable visual problems was usually in the later teens. They did not have balance problems and they were usually users of spoken English. Usher type 3 was discovered later, and is the rarest kind, in the U.K. and the hearing impairment and the visual impairment are progressive, and usually emerge in early middle age (e.g. Miner 1995, Sense website) .

However, Cohen et al (2007) write that new findings ‘challenge the assumption of distinct genetic causes underlying each type’ (pg 90). Their review outlines that visual symptoms may not occur earlier in Usher type 1 patients, simply that they are identified earlier, that not all people with Usher 1 have damaged vestibular systems and that in both type 2 and type 3 there is progressive hearing loss. Before the labelling of the different subtypes they conclude that there are ‘atypical and overlapping forms’ of Usher.

In our report we also found that the traditional picture of Usher syndrome is changing. Genetic research found genes which could be identified and linked these to a type of Usher; this meant that then the diagnosis of

the type of Usher could be made on the basis of genetic testing rather than symptom progression, which led in turn to a typology which does not match this traditional picture.

3.3 Research questions

In beginning this project our key focus was on the lives of people with Usher as seen by themselves; how the effect of the changes inherent in Usher and how knowing that they have Usher has an impact on their lives, the way they lead their lives and their concepts of themselves.

The research was intended to answer the questions;

- What do people with Usher think about having Usher syndrome?
- How much control over their lives do people with Usher have, and who is it shared with?
- What limitations are due to having Usher syndrome?
- What are the positives and achievements of people with Usher?
- What is the effect of change on the lives of people with Usher?
- Was what has happened in the lives of people with Usher what they expected to happen?
- What do people with Usher remember of their diagnosis and what impact did it have on them?

The research was then focused on interviewing the largest sample to date for a qualitative social study of Usher syndrome; a cohort of 40 was to be sought, broadly representing people with Usher 1, 2 and 3, and from the ages of 14 to 60. There is more detail in the methodology section – section 5.

3.4 Outline

In this project report we have been able to give only some of the data gathered from a two year intensive inquiry into the lives of people with Usher syndrome. This took researchers over the country, England, Wales, Scotland and Northern Ireland, talking to people from 14 to 59, with Usher 1,2 and 3. During this time, we have gathered hours of data from the voices and discussions of people with Usher syndrome who talked about themselves in answering questions. Some of those we contacted did have links with Sense or were involved support networks—others were not.

In this report we have first of all reviewed the literature relating to Usher syndrome, including the interesting and valuable ‘grey literature’ – the accounts of people with Usher syndrome about themselves. Then we explain the way in which we planned and carried out the project, the ethics and the choice of questions. This includes a look at the demographics of the respondents, where they came from, how old they are and so on. In a series of chapters following that, we look at some of the key themes emerging from the discussion with people with Usher - diagnosis, communication, mobility, family, friends and support networks, education, employment, and leisure activities. We then explore the participants self image and self esteem. We end with a discussion which includes our key points – or rather the key points which we think come from what people with Usher said.

4 Literature Review

4.1 Usher syndrome

Usher syndrome is a genetic condition which causes vision and hearing loss to various degrees, of a progressive nature. It was described initially by Dr von Graefe in Berlin in 1858 but the name comes from Dr Charles Usher in the UK in 1914 (Millan et al 2011).

It is a genetic recessive condition, meaning that it runs in families and so siblings and parents/children with Usher are not uncommon.

The predominant features of the syndrome are sensori-neural deafness and progressive vision loss due to Retinitis Pigmentosa. Three types are usually described (though subtypes exist) – Usher 1, Usher 2 and Usher 3 (e.g Bitner Glindzicz and Saihan 2014, Dammeyer 2012a). Typically the three types are used to differentiate the severity of deafness (Usher 1 connoting severe-profound deafness, Usher 2 moderate-severe deafness) the age of onset of vision difficulty (Usher 1 in early teenage years, Usher 2 in later teenage and Usher 3 in middle age). Usher 3 is then described as including progressive deafness (not so for Usher 1 or 2) and Usher 1 and sometimes Usher 3 includes balance difficulties (Millan et al 2011). It is unclear exactly what proportion of the population might be expected to have Usher Syndrome, though Hope et al in 1997 suggested it might be as high as 6.2 per 100,000, and Sense suggest as high as 15 per 100,000 (. It may affect as many as 5% of those who are recognised as deaf in childhood (who are, probably, most likely to have Usher 1 or 2) (Kimberling et al 2010, Dammeyer 2012b))

Retinitis Pigmentosa is a gradually developing disease of the eye, which begins with loss of peripheral vision (to the rod cells) causing a limited visual field and nightblindness, but for people with Usher Syndrome, it usually continues to an eventual deterioration also of central vision (cone cells), perhaps in later life (Millan et al 2011). Côté, Dubé, St-

Onge and Beauregard (2013 pg 140) talking about Usher 2, suggest that the visual impairment “may progress slowly or quickly, be stable, or degenerate until the person is completely blind.

4.1.1 Medical research and Usher syndrome

There is a considerable body of research literature on Usher Syndrome from a medical and bio-chemical perspective, all of which is outside the scope of this study. In very broad terms much of it is related to

- examining the deterioration of the retina,
- defining the exact anomalies in the genes identified
- working with other ‘cilia’ (hair cell) disorders

Some of this work could eventually lead to treatments for the eye condition (and possibly for the hearing loss). Gene therapies and stem cell implants are a goal for some (e.g. The Coalition for Usher Syndrome Research www.usher-syndrome.org) and other possible treatments include both drug regimes and retinal implants (Coalition for Usher Syndrome Research). The Coalition maintains a website which a wide range of research, links to papers and abstracts and discussion of possible treatment options (www.usher-syndrome.org).

4.2 Impact of Usher syndrome

In beginning our research into Usher Syndrome we considered first what we might expect to find, in relation to previous research about Usher and about other circumstances with similar perspectives; for example, in relation to vision loss without deafness.

Network 1000 (Douglas, Corcoran and Pavey 2006, 2009) looked for instance, at work, finance, independence, travel, technology, education and leisure. Of the approximately 600 visually impaired participants of working age, 34% described themselves as employed, 34% as sick/disabled or retired, 22% as unemployed, 7% as looking after a family. While most of those of working age got out of their homes regularly (67% left their home every day) many felt that there were barriers to mobility. A wide range of leisure activities were reported,

including watching/listening to radio, TV and music, to video or CD (about 90% of people).

Godlieve, et al (2005) reported on 93 people with Usher answering a range of questions about the impact of vision and hearing problems on independence, communication and information. Those with Usher 1 had more difficulty than those with Usher 2 in leading independent lives, as measured by the questionnaire. The degree of hearing loss was a possible explanation.

In terms of comparators with Usher syndrome, we looked at some other diagnoses and the impact they had on individuals. Of the range possible, we looked at those which had an impact on daily life, and those in which people might worry about the future. For instance, Palladino et al (2013) write about how for the diagnosis of diabetes,

The demands of managing a complicated illness like diabetes must be integrated into the normative changes in vocation, education, relationships, and living situations that accompany emerging adulthood (pg 507)

This seemed to be a similar picture to those learning to live with Usher. We also looked at research about cancer and cystic fibrosis, because these have life long effects; they are not 'over'. Unlike all of these of course there is no treatment for Usher, so it does not involve getting involved in medication for instance. It is not life threatening like cancer and cystic fibrosis. Nevertheless, the insights from these conditions give some indication of how others feel in these situations.

Thompson et al (2013) writing about young people (15-25) with cancer demonstrate that professionals often overestimate their concerns with issues such as survival and treatment, and underestimate their concerns about how they will get on with their lives – can they go to the party during the treatment regime for instance. Similarly Harning et al (2013) showed that young people with cystic fibrosis (in Stockholm) were not very often depressed or anxious – although some were both, while Havermans et al (in Belgium) showed that adults with Cystic Fibrosis were not significantly more depressed or anxious than their peers. Larsson et al (2010) showed that initially a diagnosis of cancer in young

people led to a higher level of depression. After 18 and 48 months, the group with cancer actually had lower levels of depression and anxiety than those without. Young adults with diabetes showed similar levels of quality of life measurements as those without on most dimensions of quality of life and life choices in a study by Palladino et al (2013). However, they did show differences in life satisfaction, life purpose (and drive for thinness).

4.2.1 Usher syndrome and mental wellbeing

However, in looking at Usher syndrome as part of deafblindness, there are indications that more difficulties might be expected. Bodsworth et al, (2011) while not studying Usher syndrome in particular, found very high levels of psychological distress among deafblind people, with almost half of respondents showing quite significant levels of distress as measured by a standardised tool. With a more liberal interpretation of the criterion for distress, this rose to 2 in 3 deafblind people as not feeling good about life and themselves. Bodsworth et al discussed the possible negative effects on mental wellbeing, and functional independence. In particular Bodsworth et al report a lack of services, or services geared to helping people with one sensory impairment by using the other – compensating for vision loss by using auditory means for instance. This is reinforced by other studies such as Miner (1995 and 1997). They also highlighted the difficulties of an additional diagnosis such as of vision loss to a person who is already deaf. However, it should be noted that 62% of the sample were over 65 years of age.

In 1994, Vermuelen and van Dijk concluded from 16 young people (mean age 13.6) with Usher syndrome in a school in the Netherlands, that they had very high levels of ego resilience, self esteem and social competence. Van Dijk, suggested that further research was needed to determine if this positive outlook and adjustment was part of Usher - something intrinsic to the syndrome (van Dijk, undated).

Wahlqvist et al (2013) administered a questionnaire to 96 individuals with Usher 2. They demonstrated in a (self reported) measure of physical and psychological health, that individuals with Usher 2 had significantly greater levels of wellbeing - reporting fatigue, depression,

thinking about suicide, and physical symptoms. Wahlqvist et al (2013 p 212) report that they showed “fatigue, inability to concentrate, being unable to accomplish things, feelings of worthlessness, and feelings of being constantly under strain” as well as unhappiness, anxiety and depression. This also is supported by Miner’s (1995, 1997) studies in which he discusses ideas of suicide as being frequent in individuals with both Usher 1 and 2. Wahlqvist says that suicide attempts in people with Usher 2 are six times higher than those in a control group. Miner, discussing Usher 1 in 1995 and Usher 2 in 1997 talks about how depression can reoccur throughout the life cycle as individuals adjust to change.

Bitner Glindzicz and Saihan (2014) suggest that Usher may be associated with psychiatric disorder, though based on rather out of date evidence (e.g. Hallgren, 1959). They note that one current theory is that

The genes which cause Usher may in some way predispose towards behavioural and mental problems (pg 246)

but there is no empirical evidence of this. The other theory they mention is that it is the stress of dual sensory loss which causes higher levels of emotional difficulty.

4.2.2 Ontological security, identity and social recognition

Miner (1995) talks about the effect of visual impairment on people with Usher 1, who then have to redefine their identity (as Deaf people) thereby losing a sense of the role they play in their families and communities. Similarly both Danermark and Möller (2008) discuss the idea of ontological security, recognised as a major factor in promoting self identity and constancy. This they say is based on a “sense of the reliability of persons and things” (p S120). They outline four aspects; trust, continuity, routines and predictability. Each of these are then shown to be diminished in the case of individuals with acquired deafblindness, of whom the major group is individuals with Usher syndrome. The ‘constant change and an ongoing adaptation to new situations’ (p S121) is especially seen as a threat to ontological security, the security of self identity. They also pick out the fact that frequent change in professionals and others who are supporting someone can

make it unnecessarily difficult for them to adjust. The importance of self confidence, self respect and self esteem is demonstrated, relating to individual's role in society, in their family and in their own eyes.

The lack of continuity and routines is also typical for people with deafblindness, especially those with Usher syndrome. There is constant change and an ongoing adaptation to new situations to emergent limitations in functioning. Once a person with deafblindness has adapted to new conditions, almost immediately the conditions start to change again. (Danermark and Möller, 2008, pg S121).

Finally, they note that there is not enough literature about people who have made successful lives with Usher syndrome, particularly (they note) in relation to those who have cochlear implants and use speech.

Ramsing, (2013) writing about children with Usher 1, suggests that they may have a range of possibilities for responding to a diagnosis; anxiety, despair, competent but waiting, stuck, or fighting their way through. She suggests that although they may initially appear to have depressive or behavioural disorders, that it can be Usher and its consequences that cause these responses in children and young people. Stress and the energy sapping effects of dealing with poor vision and hearing may mean that they have difficulty in learning, social and psycho-social development.

4.3 The voices of people with Usher through research

Alongside the studies relating to the condition of Usher syndrome and those that look at the syndrome as a whole, there are a number of studies which look at the lives of people with Usher more as a whole. These studies bring the whole lives of people with Usher more into focus. Amongst these studies we have in particular picked out Schneider (2006) *Becoming deafblind; negotiating a place in a hostile world*, Oleson and Jansbøl (2005) *The Nordic project (experiences from people with deafblindness)* and Gullacksen, Göransson, Rönnblom, Koppen and Jørgensen (2011) *Life adjustment*.

4.3.1 Life adjustment (Gullacksen et al)

Gullacksen et al interviewed 15 people from three Nordic countries living with deafblindness (most of whom had Usher syndrome) in relation to the 'Life adjustment' model. This postulates a progress through life from surviving to exploring, then stabilising and finally maintaining 'Living with' deafblindness. Their interviewees were all aged 25-65. In reflecting on their childhood and adolescence, while noting a number of difficulties; who to talk to about their condition, feeling different from other young people, particular difficulties with schools sports and games, and not being able to drive, for instance, overall 'most of them describe a good life' (pg. 38). The authors discuss the participants' feelings of loneliness in relation to knowing who to speak to, and vulnerability, as they were bullied or excluded. On the positive side, they speak about security within family relationships, and the benefits of seeing life 'as a challenge'. Within the adjustment model, they acknowledge that while diagnosis is often a triggering moment for readjustment, for these young participants as young people, it often was not, because nothing had yet begun to change.

Moving on from this, they describe young people as holding on to 'normality' and then a trigger, which can be a simple realisation that something is not the same again, leading into a period when they have to readjust, and that this causes a time of anxiety and disruption. One particularly potent occasion is when a driving licence is not renewed or must be returned. From this point, people realise, Gullacksen et al suggest, that they have to change.

The next stage is often grief and worry for what has been and what is to come. This is followed by trying to adjust to life with new difficulties, perhaps trying to learn new means of communication, or contact new support networks. This can be a difficult time, as Gullacksen et al suggest "apart from emotional preparedness, time, strength, and energy is also required" (p 50).

The third stage, they suggest, is finding a basis on which to continue. This might be an acceptance of being deafblind, and integrating this into a forward moving journey again. The final stage, maintaining, is reached

when participants begin to live as deafblind people, with the support they get and keeping in touch with life the way they want to. For the people in their study, Gullacksen et al say that they were mostly at the level of having already achieved this stage.

4.3.2 Becoming deafblind; negotiating a place in a hostile world (Schneider)

Schneider's work (in Australia) is based on interviews with eight people, three of whom had Usher syndrome, two people had type 2 and one person type 1.

Through the eyes of the people she studied, she examines a range of issues, looking at adjustment to a world in which it is difficult to live as a deafblind person. She looks at a range of themes in relation to her participants; being like other people, using strategies to live differently, dealing with uncertainty and change, advocating for themselves, and negotiating for support.

The people with Usher talked about accepting Usher over time – for Sam (type 2 and aged 47) only after he had exhausted every hope for a cure. Joseph (type 2 and aged 50) said that few professionals understood the needs of a deafblind person – he gives the example of an audiologist not understanding why very small hearing aids, which Joseph could not see when he dropped them, might not be the best option for him.

All three gave examples of difficulties they had had. Joseph felt very isolated and indeed for some time hid the fact that he was deafblind. He felt that everything in this life was dependent on others – his partner in particular. Joseph, and to some extent Sam, put on a face to the world which was not necessarily what they felt. Usher was something that had happened to them. Glen however, (type 1 and aged 25) deliberately decided to seek out other people with Usher in order to make contacts – and set up mutual support groups.

While both Joseph and Sam struggled with accepting Usher, Glen seemed to embrace it more; through his contacts he found that Usher

did not have to be the end of everything he had hoped for “They helped me understand my life is not limited just because I have Usher. I can get married. I can have a full life. I realised other people have Usher and have set themselves up” (p 142).

4.3.3 Experiences from people with deafblindness (Oleson and Jansbøl)

Oleson and Jansbøl interviewed 20 people with Usher from across the Nordic countries (Norway, Denmark, Sweden, and Iceland) from 17 years old to 63. 8 had Usher 1 and 12 had Usher 2. This was a longitudinal study over 5 years. Through yearly interviews as people lived with and adjusted to change they looked at how individuals constructed their own identities and, managed their daily life. They look at ‘coping’ – bearing a situation and preparing to live through changes.

Over the five years, 9 of the 20 said their vision had got worse – and in 7 of these the loss was measurable. 3 had lost some hearing.

The older participants particularly talked about how they wondered what was wrong with their vision before they knew they had Usher; younger participants had known since they were children. Many talked about their shock when Usher was diagnosed and a feeling of being alone. Some coped with the implications of the diagnosis by ignoring it – or not understanding and not wanting to know. Others accepted it and moved into the world of deafblindness – clubs and support groups. In talking about support, they discussed managing the professionals who supported them – who sometimes wanted to give them things they did not want, or who seemed to talk over their lives. They emphasised the importance of emotional support – companionship with helpers – sometimes even over help with practical issues. Their experience of being involved in the community differed; some of them felt they had to negotiate this even within their families. Involvement in the community is hard work and practical activities such as keeping house take up all their energy.

While most of their participants had achieved a good level of education, they were not all happy with the level they had achieved. Not all educational institutions had made allowances for the difficulty in vision

and hearing. In relation to work, only four of the 20 were employed in a mainstream environment, (two were in education and some were retired). 4 others worked in an environment with understanding of disability, or with other disabled people.

The people who were interviewed generally said that they found reflecting on their lives valuable. The yearly interviews gave them an opportunity to reflect, which was for some of them cathartic.

4.4 People with Usher speaking about themselves

Finally, we looked at how people with Usher had spoken about themselves, not through structured research, but with their own voices, sometimes through published books or articles, but more often through social media, through internet articles, YouTube videos, blogs and other more informal accounts. The most interesting sounding one was John E Smoke of the Flesh Eating Foundation (an electro-zombie-punk band whose singer has Usher). While his live interview is no longer visible, a transcript of an interview remains available in which he says

“At some point you'll see me trip up, nearly fall off the stage, cut my head on the mic stand, or accidentally stamp on something, because I'm blind. I'm a bit of a hazard. I'm deaf too. I'm really in the wrong job. All part of the experience though”. (John E Smoke at fearnetwork.com)

While these are not of course in any sense a representative group, they do demonstrate the lived experience of people with Usher as they feel their lives and talk about what is happening for them.

There is a huge range of such material. We chose material which interested us because

- a) it represented a different source or voice (e.g. someone with Usher 1, an article in a newspaper)
- b) it raised new or interesting points
- c) it was something which others used or referred to

As this literature is disparate and to some extent undefined, some of the key sources are listed here;

- Molly Watt's YouTube videos and blog (Molly is a young person with Usher 2)
- James Clarke's YouTube videos (James is an adult BSL user with Usher 1)
- Rebecca Atkinson's articles in the Guardian newspaper; also Robert Halford's which appeared just as we were starting this project
- Tamsin Wengraf's article in the Mirror newspaper
- Entries by Nick Sturley, Christina Hartmann and one of the 'secret deafie' articles in Limping Chicken
- The blog 'Diary of a deafblind girl'
- Hartmann's internet articles (including in the Limping Chicken)
- Accounts in Talking Sense, and other journals
- Cohen's account of Rebecca Alexander in the New York Times
- Cyril Axelrod's book 'And the Journey Begins'
- Conference papers from the ADBN networks and CAUSE in Europe (presented by people with Usher).
- Articles about or by people with Usher in *Talking Sense* - the publication of the voluntary organisation, Sense.

Obviously in these accounts from people themselves about their lives there is no common theme and no easy summary. The only unifying points are that they have Usher and that they want to write/talk about it. Within them people talk about a range of feelings and approaches to Usher. Many talk about initial shock and depression (e.g. Palmer, told by Taylor, 2004, Axelrod 2005, Henderson, 2000) or about withdrawing (Hartmann) or pretending it was not happening (Pollington, 2004). For Shannon (Fassler 2010), it is others who do not realise that she cannot see, as her eyes appear normal – and then they think she is being rude. The prospect of impending blindness worries most of the authors - some of whom have been told they will 'go blind'. The effects of not being able to see while already d/Deaf are mentioned;

It's really, really complicated and devastating, especially as a deaf person. It's taking away something that you depend on the most in your every day life. (Hartmann no date).

Hartmann also talks about how losing her vision (she was already deaf) changed her relationship with her partner; he was afraid she would change, she was also afraid that she might change into someone he would not like (in fact, when the article was published, they were still together)

Our particular challenge may be unique, but the fact that we face them isn't. If you're with someone longer than a year, the odds are that you'll encounter some kind of major change. Maybe it's a job transfer, childbirth, or a death in the family. In that sense, my story isn't anything special.

Atkinson talks about grabbing opportunities while she could still see (2004). However, most of the accounts then go on to dealing with the outcomes, as Dainton (2007) says:

I have learnt and still learning that it's better not to assume the worst scenarios, not to think about the worst but the better aspects of things and people.... I find taking on these more positive attitudes really helps me to cope with difficult situations. Sometimes it's best not to ponder, not to think at all, but just relax.

Deafblind Girl (2011-12) says:

After being diagnosed with Usher, I figured there are a few things I could do 1- I could ignore it, and pretend that I don't have it. 2- I could huddle up in a corner and let it run my life or let it take life away from me. Or 3 - go on full tilt, find out all the information that I could and try to live a full and happy life.

She, like Halford, (2012) chose the third option – Halford says “So many wonderful things have happened since my diagnosis and now, age 50, I feel that I am living a second life. ”It is often the fear and anticipation which brings more worry than the actual living with Usher. Hartmann hid

her diagnosis, and Secret Deafie says, “The fear was far more crippling than the reality. Bring it on- at least then I am dealing with it, rather than worrying about it.”

Some say that using a cane is such a sign of visual impairment that they cannot bring themselves to do it – while Clarke (2011) shows himself with, and without, a cane, getting through a station, to demonstrate how he has adapted and how much of an improvement it is.

Some give advice to others, either others with Usher or those who are alongside them. Watt gives advice to teachers on how to help pupils who have poor vision and hearing; Sturley (2012) tells Deaf people that they should not assume that people with Usher are unable to look after themselves, - he says he would tell hearing people the same, but he expects that Deaf people might know better.

Of course, it might be expected that those who write, speak or sign about Usher would be those who have already come to terms with Usher. While it cannot be said that these voices are a complete representation of Usher syndrome, they are the words of people with Usher on their own terms.

4.5 Moving on the next steps

From the literature, therefore, we identified a number of key themes we wished to investigate and some ideas we wanted to explore. We had planned a project which involved interviewing more people with Usher than any project before (40) and we hoped to find people with Usher 3, who were absent in the major interview studies. We wanted to find out how people with Usher actually lived their lives and what their concerns were. Was Usher central to their current life and thinking or was it only peripheral? What factors in Usher had the biggest effect on their lives?

There were indications from the literature that stress, anxiety and depression might be the prevailing stories from the world of people with Usher. Was this so? From hearing what those who had already spoken had said, we wanted also to hear the voices of those who had

not spoken. This would include young people (possibly with their parents) as they are now, rather than retrospectively, and people at different stages of Usher and vision loss, with different experiences, across Britain.

5 Methods

5.1 Data collection methods

In this chapter we explain why we chose the methods we did for collecting the material about people with Usher syndrome and how successful we feel these were. We also outline some details of the participants in the project.

Our work was based on a case study methodology, that is, that we looked through the lives of individuals to gather exploratory information about the way in which they, each as an individual, responded to the challenge of Usher syndrome. We gathered qualitative data, from which we can build a picture of the lives of people with Usher. This methodology is not intended to be at all *representative* in the sense that the participants were not chosen to represent and be typical of the population. However, it allows the research to show a full picture of a range of different responses and to explore through these the implications for people with Usher syndrome. Thomas (2011) describes case studies as a way of gathering data in a more multi-dimensional way. Although each study is of one person, through looking at a range of people, a number of different aspects can be explored. Stake (1995) discusses case studies as individual or instrumental – where the individual is the main focus or where individual studies illustrate the main points. We looked at individuals to highlight similarities and differences, a comparative case study, which Schwandt (2001) calls ‘cross case analysis’. Our case studies stand alone, as each individual describes their own experience as a person with Usher but also together, as they create a multi-faceted picture of the way people with Usher conceptualise themselves and deal with life.

5.1.1 Interviews

We chose to use interviews as the main method of gathering information about the participants. These were semi-structured, extended interviews which encouraged people to talk about their lives. Interviews provided us with an opportunity to gather certain kinds of data but also allowed

people to say what they wanted. As Denscombe (2003) says, the interviewer was able to be attentive, sensitive, non-judgemental, adept at using prompts and checking to avoid misunderstanding. Within the interview structure we were able to move to questions which the participants wanted to talk about, to use additional questions and to leave some out. For example issues which participants found too sensitive could be left out.

Interviewing people with Usher was also the right choice for a number of other reasons. We wanted to be able to communicate with people who had hearing and visual impairment – using writing (questionnaires) would have been problematic. We also wanted to know not just simple answers but expansive ones, to explore feelings which people would be unlikely to share on paper. We knew that for some participants English would not be a first language. To provide them with equal access to the project, we ensured that they were provided with interpretation into BSL and SSE (Sign Supported English – the use of signed vocabulary in an English word order).

The interviewer had a good knowledge of BSL and made social contacts and followed conversations. However we were very aware that people who have difficulty with vision use a variety of sometimes quite subtle adaptations to sign language. For this reason, we chose to support them with an interpreter of their choice, someone who would know to what extent to restrict the visual frame of signing, to know how to use hands on signing if necessary, to understand lighting, clothing and other restrictions for people with Usher (Field 2011). Almost all of the 11 participants who used a BSL/SSE interpreter used someone who knew them very well and could adapt in this way.

Where interpreters have voiced the words of a Deaf person using BSL/BSL with SSE, we have added the letter I in brackets, in bold, (**I**) after the transcription. This is a translation of sign – BSL or SSE into spoken language by an interpreter.

Through using face to face interviews, we were able to gather subtle information about how people felt about aspects of Usher (Thomas

2009). Expressions, tones of voice and pauses all gave an impression of how people felt about what they were saying.

Originally we intended to interview 40 people, and to interview 10 of the younger age group twice, once at the beginning and once at the end of the project. In fact we interviewed 42 people, and two more joined focus groups although they were not part of the original interviews.

Interviews were recorded and transcripts made for analysis. Nearly all of those who were interviewed in BSL/SSE were recorded on video, along with the interpreters, and then transcribed. Where signing participants had opted not to be videoed, or chosen to be interviewed in a public space, such as a café, the interview was only recorded using a voice recorder

5.1.2 Focus groups

As detailed above we had initially planned to interview 10 people from the young person's group twice. This was partly due to the fact that we thought that we would be able to recruit these people more easily so that we would be able to give a gap of at least 12 months between interviews, and also due to our focus on change we thought that the young people would be encountering more changes on a regular basis than the people in the other two groups. However after the initial stage of the research we realised that this would not work; young people were not the most expansive about their thoughts and they also were not in fact the most easily recruited. We therefore decided to adapt the project and carry out two focus group interviews – one with two participants using spoken English and one with three participants using BSL. Two members of the BSL group were new to the project. We also interviewed three of the younger age group for a second time. For the focus groups and the second interviews, we used a very different format, giving a range of discussion items, relating to Usher, asking them to reflect on different scenarios, rather than asking them to talk about themselves. They were asked for example what advice they would give to someone whose family all think he should use a cane, when he doesn't want to, or to say whether life events such as graduating, deciding to use a cane, a parent dying, or telling a partner they have

Usher had a big impact, some impact or no impact on their lives. For the young people, they were asked questions with similar concepts but did not have the opportunity to reflect on them together as we were unsuccessful at trying to get a group of young people together.

In interviews with the young people, parents frequently sat with them and also made comments. Parents were also asked for permission to use their contributions, where appropriate, as on occasion they provided information which the young person did not (though of course, this is **not** the voice of the young person themselves.)

5.1.3 Self Image Profile (SIP)

As well as taking part in a face to face interview, participants were requested to complete a self image profile (SIP) (Butler, 2001; Butler and Gasson, 2004). The SIP is a standardised measure of self image and self esteem. It is delivered usually as a paper exercise in which people indicate on a scale from 0-6 how much they think they are like certain characteristics (happy, fit, confident) and then how much they would like to be those things. There were separate questions for those aged 17 and under, and those aged 18 and over

There is more detail about why we chose the SIP Section 13. However, we were pleased to find a tool which

- Focused mostly on the positive rather than negative aspects of life
- Was fairly easily converted to other formats (including braille) and to BSL – it did not require wordy translation and explanation
- Was standardised – although not on the d/Deaf population (and was not delivered in a standardised way to our groups)

The SIP proved to be an interesting measure in itself and gave people a sense of satisfaction. Even those who did not say much in the interview enjoyed filling it in and thinking through the issues.

5.2 Recruitment and Participants

5.2.1 Recruitment

Originally we had planned to organise the participants into 3 age groups – people aged about 15 years old, about 25 years old and about 45 years old. We chose these age categories as we thought that people within these age groups would be negotiating significant life events, for example career choices, relationships, and parenthood. Of course people are not all the same and not everybody would be experiencing exactly the same things in each cohort, but we felt that it would provide a useful starting point in exploring the lives of people with Usher syndrome

As we began the recruitment process we realised that we could not recruit the numbers of participants required to adhere so rigidly to the categories of 15, 25, and 45 years old so we opened up the age bands to include young people aged 14-20 years old, younger adults for people aged 21-35 years old, and older adults for people aged 36-56 years old. We felt that this provided a balance between maintaining a focus around life course events, and allowing a number of people to take part in the project. Due to the sensitivities of the research it was felt inappropriate to conduct the research with people under the age of 14. Furthermore all participants had to know they had Usher.

We had hoped to interview 15 people aged about 15, 15 people aged about 25, and 10 people aged about 45. We thought that the people aged about 15 would be the easiest to recruit through the contacts that were available to us. In fact it was the older adults who were the easiest to get in contact with; this may have been that they felt that they had more experiences to share or greater time to reflect on their thoughts and opinions of Usher. Because of the way in which we recruited participants, younger adults who were not in contact with other people with Usher, or not seeking services may have been excluded from taking part in the research. However as will be shown below we purposefully tried to engage people with varying degrees of involvement; such as in Usher groups or activities, or contact with Sense. Participants were recruited to the project via a variety of means:

- Advertisement in Talking Sense
- Advertisement in the RP Fighting Blindness Newsletter
- Advertisement in Deafblind UK's newsletter
- Snowballing – where a person who had taken part in the research (or a person close to them) had contacted another person known to them with Usher and told them about the project.
- Advertising on Facebook on Usher related groups
- Social Services
- Teachers of the Deaf
- Email list of BSL interpreters
- Teachers of VI pupils
- University of Birmingham website
- Professional contacts
- Flyers at conferences
- Advertisement in the Ear Foundation newsletter

The most successful means was through social media – through our material on websites and through personal contacts on Facebook groups about Usher.

A number of people got in contact through the website RP Fighting Blindness, some of whom were too old for our research – we had decided not to look at people of retirement age. The fact that this group, rather than Sense, appeared to be an initial point of contact for some people may indicate that they see themselves as visually impaired rather than as having dual sensory impairment. An area of further research could be with people who are past retirement age and their experiences of Usher.

5.2.2 Participants

We interviewed 42 people with Usher syndrome. Of these 16 were male and 26 female. Their ages ranged from 14-56 years old. There were 12 people aged 14-20 in the young people's group, 15 aged 21-35 in the younger adults group, and 15 in the older adults' group, aged 36-56.

Two people used hands on BSL, seven people used BSL, and two people used BSL with additional SSE (sign supported English) elements. The remaining 31 people used spoken English.

We also recorded their ethnicity; 30 described themselves as white British, and the others came from a range of ethnic backgrounds, recorded in the table below.

We also asked about the type of Usher with which participants were diagnosed. Of these, 15 had Usher type 1, 22 type 2 and 5 type 3. We were very glad to have some people with type 3 as none of the cohort studies we had found (as mentioned in the literature review) had included anyone with type 3.

We were keen to follow up family relationships and to find brother/sister pairs, relationship partners who both had Usher and parents and children who both had Usher. We were unable to find a participant parent/child group and no one in the study had a parent or a child with Usher. However, we interviewed one couple both of whom have Usher, and three sibling groups (two brothers and two pairs of sisters). Furthermore 4 people had a sibling with Usher, and a further two people had two siblings with Usher. One person had cousins and other family members who also have Usher.

The geographical spread of participants was great, and people lived in various parts of the country. We also included one person with Usher who was from the Republic of Ireland as she was in a relationship with someone who also has Usher, and we wanted to explore some of the experiences of being a part of a couple where both partners have Usher. Of the remaining 41 people, 3 were from Scotland, one from Wales, 1 from Northern Ireland, and 36 from England. The spread of people across English regions is in the table below.

Category	Number
Male	16
Female	26
14-20 (young people)	12
21-35 (younger adults)	15
36-56 (older adults)	15
BSL	7
Hands on BSL	2
BSL/SSE	2
Spoken English	31
White British	30
White Scottish	3
British Asian	3
African, Black British, White Irish, White Welsh, Chinese, no answer	1 each (6)
Usher type 1	15
Usher type 2	22
Usher type 3	5
Greater London	7
North West	6
South East	6
East Midlands	4
Yorkshire and the Humber	4
South West	3
Eastern Region	3
West Midlands	2
North East	1
	Total 36
Scotland	3
Wales	1
Northern Ireland	1
Republic of Ireland	1

The two people who were part of the BSL focus group but not part of the first round of interviewing were both females, aged 50 and 59, they came from the south-east, and described themselves as white British. One had type 1 Usher, and the other had been told that she had type 1 Usher, but was waiting for the results back from some genetic testing as it was believed that she might have type 3 Usher, as she had been born hearing. One of these people was the sister of somebody whom we had already interviewed.

5.3 Ethics

Ethical approval for the research project was obtained by the University of Birmingham's Ethics Committee and ADASS (The Association of Directors of Adult Social Services).

It is recognised by the researchers that talking about Usher syndrome and its effects on individuals' lives and experiences may be difficult and upsetting. Participants were reminded that they did not have to answer any questions that they did not wish to answer, and that they could stop at any time.

If during the interview, or after on reflection, the researcher felt that the participant might benefit from contact with particular support groups contact details were made available for agencies such as Sense, Deafblind UK, the Samaritans, and Citizens Advice.

One of the disadvantages of a small sample of participants from a small population is the potential for other people to recognise the participants in the outcomes of the project. The participants were made aware that other people may be able to recognise them, although we have omitted or changed identifiers where appropriate e.g. home towns, schools, workplaces. All the names of people in this report are pseudonyms.

It was important to get informed consent from participants. Participants were provided with an introductory letter from the research team, a more detailed document about the project, and a consent form which potential participants were encouraged to send back in the prepaid envelope provided. Generally the information was provided in standard print through the post, but where requested information was also provided in

braille, electronically, on CD, in large print, and on coloured paper. The young people were provided with an information pack containing an introductory letter, information sheet, and consent form. An information pack was provided for their parents (who also had to consent for their son/daughter to take part in the project).

In order to thank the participants for giving up their time they received a £20 supermarket voucher.

We did not want to keep the contact details of potential participants so where appropriate an information pack containing these documents was passed to a third party (known to both us and the participants). These people were useful in providing further information to participants to encourage them to take part.

Before contacting any potential participants the interview schedule, consent forms, introductory letters, and project information sheets were examined by the University of Birmingham's Ethics Committee. These were approved as appropriate for this study.

Knowledge that a person had Usher (and its implications) was a criterion for taking part in the research.

5.4 Analysis and reporting

The interviews (audio or video files) were transcribed and a Word document produced. This was then imported into NVivo which is a software package used by researchers to help analyse qualitative data. Using NVivo the data was coded into broad themes. Although driven by the questions asked in the interviews these themes emerged from the data.

After this initial stage of thematic analysis the data within specific fields was examined to draw out key ideas which are common or areas of interest. The data was then grouped under these nodes of interest, using quotations from the transcripts. From these, we looked for recurring issues, for unexpected responses and for areas in which there was broad agreement. We then looked across these issues to find out what was common between the areas and what was different in the

areas we had identified. From this we developed our reporting using quotations from individuals to support the discussion.

At the time of writing we have already reported some of the findings in a web based report and at a conference. We will intend to continue to report on the website, in academic journals and in specialist magazines, and on the web, all with the kind permission of Sense.

5.5 The success of our methods

5.5.1 Representativeness

The methods we had chosen to collect our data worked and we were very glad to have had the opportunity to have interviewed 42 people with Usher and involved 44 people with Usher with our study.

We are of course very aware of the limitations. We did not intend to gain a representative sample and we cannot claim that all life (with Usher) is here. Natural biases were built into our data. We not have any young people who did not yet know they had Usher, which means we do not know how young people worrying about their vision might feel –though we have had some older people discuss how they felt when they were young. Our calls for participants went mostly to people who were interested and involved – and certainly did not go to anyone who did not want to acknowledge their Usher at all. We have not found those who are the most isolated, the most depressed, who will not have been in contact with our advertisements or who did not want to speak to anyone. The colours they would have added to the picture would have changed it. We did recruit a number of BSL users but we did not, despite our efforts, recruit someone who used fingerspelling, and only two who used hands on sign language. While we did recruit brother and sister pairs and a couple, we did not find a parent/child pair

However, we did try hard to find people from a range of ethnic backgrounds, and we did – we also tried to find people who were not in regular touch with Sense (to get this perspective) and we also found these. We are pleased that during the project we interviewed or spoke to 44 people with Usher syndrome; possibly making this the largest cohort of people with Usher to be studied in social research of this kind.

While not making it representative, of course, this meant that we were able to explore a number of different ideas and get some impression of key features across a range of people. We were pleased to recruit 5 people diagnosed with, or believed to have, Usher 3, a group which has been very little represented in literature before. Even so, this was a very small number and we could not hope to make general statements from this number of participants. In addition, we could not be sure that these people would not have been diagnosed differently in earlier times, before genetic testing – perhaps indeed other studies, where people had not had genetic testing – also, unawares, contained people with Usher 3.

5.5.2 Other issues

We were disappointed not to be able to make the focus groups bigger and to include the young people in them. People with Usher talking to each other about Usher was useful for us, as researchers, and to some extent useful for the people who came.

For the researchers, this was a fascinating insight into the lives of people with Usher – through visiting people in their own situations – homes, workplaces, or in other places of their choosing. It also meant that we were able to see people's feelings and emotions as well as just their words.

6 Diagnosis

The central theme of this chapter is diagnosis, and the impact of diagnosis on the lives and experiences of people with Usher syndrome. In considering the theme of diagnosis we see that the impact of diagnosis stretches far beyond the day on which an individual is told that he or she has Usher. We shall explore the experiences of participants with regards to their initial diagnosis including reactions on diagnosis, young people and diagnosis, the importance of information, and the additional medical conditions that might also accompany Usher syndrome. Following this we shall survey how developments in the genetic analysis of Usher blur the traditional understandings of Usher types and the impact that this has on the lives and experiences of people with Usher. We shall then look at participants' experiences of living with a diagnosis of Usher, with particular reference to accepting Usher and the adjustments required because of Usher. Finally in the discussion section we shall suggest that the three themes of change, predictability/uncertainty, and diversity may provide the most appropriate lens through which to understand the lives and experiences of people with Usher syndrome.

6.1 Initial diagnosis

The majority of the adult participants spoke with emotion on the subject of their diagnosis of Usher. They spoke about the harsh way in which they were told that they had Usher:

I went to the hospital. Polish doctor. You have RP. That's it. You're going blind. Goodbye. That was it! Not a good experience. (Dave, 46, type 3.)

I went to the doctors, and they referred me to the hospital. They said you've got RP . straight off. that's it. and my world completely crashed down. **(I)** (Gill, 59, type 1)

I was just going for a normal, routine, eye test, check up, and as I said the optician asked if I'd damaged my eye. Referred me on to the clinic. And they tested my eyes, and then next thing they told me was 'sorry you're going blind.' I wasn't expecting that at all. And I think it was the shock. I could see their mouth going but it wasn't sinking in. I came home and I was in bits. I guess that's normal I suppose. I was in bits. I couldn't even tell my mum what it was. She had to then ring the clinic to find out. It was just the way they'd done it, sorry you're going blind. And that was it. (Alice, 41, type 2.)

Experiences like Alice's were common in that participants had gone for a routine optician's appointment, had been told that they had a speck of something at the back of the eye and had then been referred to the specialist. Although very shocked by their diagnosis a number of participants reflected upon the fact that a diagnosis made sense – for example the times when they had tripped over things that had been obvious to other people, not being able to catch balls like their friends, and near misses whilst driving.

Although shock and disbelief at the diagnosis of Usher, for Nicola (23, type 2) there was relief that there was nothing more serious as mitochondrial disease had been mentioned, and for Kat's (17, type 1) mother there was relief to finally get an accurate diagnosis, especially after Kat had previously been diagnosed as having Waardenburg syndrome. For Rob (22, type 1/2) finally getting a diagnosis of Usher exonerated his mother from accusations of child abuse, due to the number of head injuries that Rob had suffered from as a child as a result of his narrowing visual field.

However a diagnosis of Usher was not unexpected for some participants. Harry's (15, type 2) and Jess's (15, type 3) mothers felt that they knew that their child had Usher before they received an official diagnosis from medical professionals. This understanding had come about by observing the difficulties that their child was having in mobility and at school combined with information that they had received from NDCS (National Deaf Children's Society) on visual impairments and d/Deaf children. Hamid (44, type 2) had diagnosed himself with Usher when he was in his early twenties. Dissatisfied that opticians and his GP did not understand his vision difficulties, through contacts that Hamid

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made at Sense, Hamid travelled to the United States and spoke to consultants there, and took this knowledge back to consultants in the UK. Despite having difficulties in coming to terms with the potential consequences of Usher, it was a source of pride for Hamid that he had diagnosed his Usher himself.

6.1.1 Reactions on diagnosis

Participants spoke of many different reactions to their diagnosis. For some, like Sally, shock and denial were their first reaction:

And the guy was och you'll be blind by this time next year. You've got RP. This is what you've got. And you're kind of shell shocked... It was denial obviously as well. Too raw to go into... (Sally, 42, type 2.)

For Dave (46, type 3) denial was also his first reaction but what we also see is that this denial cannot remain forever, "I tended to shut it away and pretend it was never going to happen. But it does." As will be discussed in section (6.3) on living with Usher, we see that due to the slow nature of change which is typical of the vision loss associated with Usher and Retinitis Pigmentosa it may be possible to ignore for a while (months or even years) after diagnosis but there comes a point when it can be ignored no more. This was also the experience of Gullacksen et al (2011) in their interviews with people with Usher.

Depressive feelings and clinical depression were also common, with 2 out of the 12 young people who took part in the in-depth interviews suggesting that they had received professional counselling. However it is interesting to note that requirement for counselling came not straightaway after diagnosis but resulted from problems and difficulties relating to Usher that arose over time. Seven out of the 30 adults who took part in the initial interviews suggested that they had developed depression as a result of their diagnosis of Usher. Some participants no longer classed themselves as depressed whereas two were continuing to take medication. As well as medication, counselling, yoga, and mindfulness were suggested by participants as ways in which they had overcome/managed their depression. Depression was a feature shown by previous researchers, including for example, Miner (1995; 1997).

Alcohol and recreational drugs had also been used by a small number of participants to help them cope with the diagnosis of Usher and the on-going implications of being a person with Usher.

For some participants, researching Usher was their way of understanding their diagnosis, especially as the majority of participants felt that they were given inappropriate information and guidance when they were first diagnosed. As the quotation below from Lucy shows, individuals' need to research can change over time, as well as highlighting the fact that people with Usher, even if they are siblings, have very different reactions to diagnosis:

I think at the time it was just the diagnosis that set me off. (Sister) was completely different – she was reading up all about it. She kept sending me news articles, and I was used like nah, nah, nah, yet now I think we're completely opposite, where I'm the one doing all the research, well not research but I'm part of some groups, read up about it, and just generally keep an open eye about it, and Megan just doesn't really care – she hasn't got a dog, she just uses an umbrella as a cane, because she doesn't want to look like she's blind. So I really I think in the end I've been able to accept it better than she has, but I think that it was my parents that struggled the most. My mum especially was very upset. (Lucy, 27, type 2).

Lucy also highlights the fact that for parents of people with Usher, finding out about their (adult¹) child's diagnosis was an upsetting and worrying time. Richard also speaks about the difficulties his mother had with accepting his diagnosis of Usher:

¹ We did not ask the young people (14-20 years old) about their parents' feelings/opinions on diagnosis.

My mother was very worried, and still is. She bears a lot of guilt. Because of my deafness when I was a baby and everything, she feels responsible for that. And I think that was a tough one for her. And the diagnosis. And she felt awful. And in the end I just had to turn round and tell her just get off because she was projecting her fears and her negativity on to me, and I was just get away I don't need that... But since I had that conversation with her, she has kind of backed off, and just let me be me. I know deep down she still worries and feels guilt, but I can't... that can't change really. (Richard, 48, type 2.)

Parents of people with Usher who had been diagnosed in childhood tended to turn to specific organisations and agencies for support and information such as NCDS, Sense, the Ear Foundation as well as online groups on social media. The importance of support and networks for individuals as well as their families will be discussed in-depth in chapter nine but is also a reoccurring theme throughout this report.

A diagnosis of Usher also provoked some more extreme reactions - Dave had a vasectomy and Lucy decided to get married:

I think that I must have panicked because I do think that I rushed into getting married! I'm very happily married, but literally we were engaged very, very quickly and I think I was "nobody's going to want me." (Lucy, 27, type 2.)

Carolyn and Nicola both said to their partners that they would understand if he no longer wanted to be with them as they did not want to 'burden' him with someone who was blind:

(Husband) was very good. We'd been married a month when I got diagnosed. I remember literally turning around to him and saying, I shouldn't have done it but for some reason I did, saying there's the door, if you wanna go, I'll let you go. I'd rather you start your new life now, while you're still young, while you still can, cos obviously I wasn't pregnant. Cos this means I need a carer and I want you to be my husband, and the father of my kids. But it also takes a lot more care than a normal wife would. So I can completely understand if he's going this is too much work, what've I done, what've I signed up for, there's the door. But he didn't. Which I'm very thankful for now. For some reason I felt that I had to do that. Give him at least the choice. If he wanted to go, he could go. Cos it is a lot to take on board. (Nicola, 23, type 2).

Nicola's husband did not leave her and instead they decided to have a child together earlier in their marriage than they had anticipated as Nicola "wanted to see the child grow up."

6.1.2 Telling young people about Usher

Of the 16 adults in the study with Usher type 2, only 4 found out about their Usher when they were under the age of 18. All bar one of the twelve adults with type 1 Usher found out about their Usher whilst at school. Two of the four adults with type 3 Usher were diagnosed whilst they were still at school. It is suggested that the people with type 1 Usher were identified as having Usher at an earlier age as they were educated, often in residential schools for Deaf children, and surrounded by people who had a greater knowledge of Usher syndrome.', Furthermore, the vision loss associated with Usher tends to begin earlier with people with type 1 Usher. This knowledge of Usher syndrome by the Deaf community meant that Rebecca (29, type 1), (whose parents had not told her about her diagnosis at six years old) at about 17 or 18 years old found that other students started to notice her nightblindness and balance difficulties and told Rebecca that she had Usher syndrome. Although Rebecca believed that her parents were acting in her best interests, she strongly suggested that parents should tell their children about their diagnosis of Usher.

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The experiences of adults who were told that they had Usher whilst at a young age provide us with adult reflections concerning the experiences of people who are told that they have Usher whilst they are young. There were particular difficulties regarding communication between SSE/BSL users and their hearing parents and medical professionals:

I thought that I was fine, that I just needed glasses to help me read. I didn't realise it was Usher. My mum had tried to explain, I'm not criticising my mum in this, she'd explained what she thought I needed to know. But obviously there weren't interpreters for doctors. there wasn't anything like that. I only realised that I could take interpreters to the doctors a few years ago actually. That's why I didn't really understand what was going on. **(I)** (Faith, 23, type 1.)

There was no interpreter. I was quite young so my mum had come with me. I didn't really know anything. I mean my mother doesn't know that much sign language, and she was talking with the doctors. And she said that I had bad eyes, and possibly I could go blind. But she didn't say anything about Ushers. They said RP, but they never said Ushers. **(I)** (Jayne, 28, type 1.)

Farah's more recent experiences however show how the appropriate communication support can help to have a positive outcome on how an individual feels about their diagnosis of Usher:

Before I didn't know what Ushers was but as time has gone on I've coped well. I've had one to ones with the sense worker, been to the doctors, with an interpreter that told me. I thought that was good I've got some support for what I have. What Ushers means. What's involved in having Ushers. What RP is. A clearer understanding and I feel good now I know what I have. **(I)** (Farah, 17, type 1.)

The majority of the young people in the project could not remember being told that they have Usher. As Harry said, "I don't, but my mum does" and Chloe and Leah's mother said, "they just don't know any different." Or if the young people could remember something it was being told that they "had an eye condition" (Bethany, 17, type 2.)

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Some parents spoke about the fears that they had in telling their child that he or she has Usher. However, as this comment from Daniel's mother shows, being told that you have Usher may not be as significant as parents imagine it to be:

It was 2 Christmases ago, and I was taking you to your panto with youth club, and we were following a motorbike, and you were really, really worried, that the cyclist on the motorbike, wouldn't be able to see where he was going because it was dark out. And I explained, that actually most people could see where they are going in the dark, and it is you and a handful of other people who can't, because you've got Ushers. And you went, oh right, don't forget to park round the corner where the car park is mum! And that was how we started talking about it. (Mother of Daniel, 15, type 2.)

The extract from the interview with Dima (15, type 1) reminds us however that although participants may not have remembered the words used or a particular day on which they were told that they had Usher, the act of being told may arouse strong emotions:

Researcher: Do you remember being told that you had Usher?

Dima: No

Researcher: Do you know how old you were?

Dima: I can't remember at all. But I had bad feelings when my parents tell me that I have Usher.

Researcher: So your parents told you that you had Usher?

Dima: Yeah.

Researcher: Can you remember how you felt?

Dima: I felt sad and shock.

Some of the adults with Usher pondered on the best time to find out about a diagnosis of Usher. Linda (46, type 1) and Doreen (53, type 1), who were diagnosed with Usher whilst at school, recognised that the teenage years are difficult and it is unfortunate to get a diagnosis of this at a particular time. This was supported by Hasan (15, type 1) who said, "I wouldn't like go into detail about Usher and stuff because I don't think at the age of 15 teenagers are really interested in that kind of thing." Dave (46, type 3) and Sally (42, type 2) in their joint interview, suggested

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that they were glad that they did not know about their diagnosis in childhood:

Dave: I don't think I would have wanted to know. Sometimes ignorance is bliss.

Sally: That's a line I've used so many times. When you're first diagnosed you don't want to know because there's nothing that you can do about it. Now I know ignorance is absolute bliss.

Vicky and Nicola however spoke about the advantages of knowing at a younger age, as younger people are seen as more amenable to change:

...but [children] are still learning, they're easily adaptable. I suppose it is easier for me, I suppose in comparison to a 40 year old, just been diagnosed, just starting to lose their sight. Cos they've had 40 years of normalness if you like, you know, I suppose the younger you are the better. The easier you adapt. The easier you can change. (Nicola, 23, type 2.)

However as we shall see in the section on living with a diagnosis of Usher, the age of diagnosis is not always significant. What is more meaningful is when a change occurs and a person has to adapt and readjust in order to continue with their life.

6.1.3 Importance of information

Participants spoke about the lack of information about Usher, particularly after their initial diagnosis. Sally commented:

I was nursing at the time, and I was saying the kind of line of work that I'm in, will that be ok? Oooh no! You'll be blind in a year! And away he went. And I was left shell shocked. No information. No organisation to get in touch. (Sally, 42, type 2).

Participants often found it difficult because they felt that medical professionals could not give them accurate information on the condition:

Cos when I asked how bad it is, he said I can't tell you. You're living with a ticking time bomb. I could send you home from here now, and tell you everything's fine, and in a week's time it could have gone. I can't predict how fast or how slow, how it is actually going to alter.

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And that's what he told us. (Mother of Leah and Chloe, 19 and 16, type 2.)

As will be shown throughout this report the progressive nature of Usher is one of the most challenging aspects of the condition, and individuals want information and guidance on how it is likely to progress. Although recognising that medical professionals may not be able to provide individuals with answers to all their questions, there are more appropriate ways of dealing with questions than in the example above. One piece of advice that participants wanted other people with Usher to know about was the specialist knowledge available at Moorfields Eye Hospital in London, and recommended that if possible individuals with Usher should get referred there. Alice (41, type 2) recognised that doctors may in fact tell you important or useful information. However the piece of information that you focus upon is 'you're going blind.'

Alice, Vicky, Brian, Julie and Sally spoke about the importance of information which can be taken away after the initial diagnosis. Brian suggested that this material be available in a variety of formats including large print. For Lucy the information that was available was not suitable for her needs at that particular time:

Frustrating! Like they told us things that were a bit unnecessary like talking books. I'm sure if I got worse, visionwise, and got the diagnosis that would be quite useful to know. There is a lot of information out there that is not really suitable for the vision loss that we've got now. Whereas there are things that we could have been told, that we've only found now. (Lucy, 27, type 2.)

Participants also highlighted a need for the availability of information as time goes by not just at the point of diagnosis as it is often not until a specific event occurs e.g. the need to use a mobility aid, or giving up driving, or a certain level of acceptance has occurred, that people are able to ask for help and advice as necessary:

You may not want to think about it. That was the main thing for me. I didn't want to think about it, I didn't want to admit it, I didn't want to realise it. So I denied it. And it wasn't 'til my sight got worse that I kind of went I've got no choice now, I'm going to **have** to make this phone call. I'm going to have to take this route. I need to, especially with

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(daughter) coming along. So yeah it's accepting it. The hardest bit, but once you've accepted it, make that move because they're not going to come up to you. They're not going to knock on your door and go hi I'm from [local sensory support team], would you like some equipment, would you like a long cane, you need to do it. Push it. You need to chase them. That's good advice. (Nicola, 23, type 2.)

A number of participants also spoke of the added complication that individuals do not know what information is available which means that they cannot ask for the appropriate information as necessary. Kat's mother explains:

I'm sure that there's probably a lot lot more out there that we could access but people don't tell you about them, so we don't know about, so you don't ask, and you can't ask a question if you don't know which question to ask can you? And I think when she was first diagnosed I was bombarded with information, you know it came from every single direction and I wasn't in a fit state of mind to take it all in. And by the time I was, we're talking a couple of years later, till I finally went ok... It was actually my mum that went to me (name) you know you need to sort yourself out because Kat's life is going to be what you make it, so need to... So then it's down to me. We're off and at'em! (Mother of Kat, 17, type 1.)

This comment from Kat's mother also echoes comments from other mothers of young people with Usher who felt that they had to work very hard to find the latest information, get the appropriate support, and obtain advice for their child.

In the absence of information from professionals, and to complement this information, participants spoke about conducting their own research on Usher. A number of older participants who had been diagnosed 20 or so years ago spoke wistfully of how it is a lot easier to find information nowadays, especially with regards to the internet and social media. Bethany's mother spoke about the importance of there being someone on the end of the phone to speak with, and for Gill it is was important to be able to access signed information rather than relying on written material. Participants also warned that it can be 'dangerous' to do your own research, either because the results can be scary, or that you don't

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know the source of information, and particularly in reference to cures and treatments for RP, could potentially make your vision worse. Dave warned about getting “obsessed” with regards to cures and treatments. Nicola shares her experiences of researching Usher:

Just don't Google it. Scares the crap out of you! Everything negative is on there. I think that was my problem, because there was nothing out there, I had to self research. And when you're feeling low and scared as it, you're naturally drawn to all the negative stuff that's on that search. And you scare yourself. And you think the worst. And then you go back to speak to your specialist, oh I've read this, I've read that, he said to me that it's very rare with Ushers that you'll lose your sight completely... (Nicola, 23, type 2.)

Not everyone was searching for cures however. As Jess (15, type 3) commented, “Don't really want one. I'm used to how I am. If there is one that's cool but it doesn't really bother me. I'm ok.”

Many of the participants in the project suggested that contact with other people with Usher is very useful, especially in the early stages of diagnosis, in order to compare experiences and outcomes with other people with Usher. Contact with other people with Usher will be discussed in more detail in chapter 9 which deal with friends, family, and social networks.

Having been diagnosed with Usher generally means that individuals are placed under the care of a consultant and, among other things, are invited to attend visual field tests and other monitoring appointments on a regular basis. A couple of participants had, however, been avoiding the appointments as they did not want to be faced with further, potentially upsetting information:

I don't worry about it every day. I don't think about it. I try not to. You've got a life to live. That's very much my attitude really (voice starts to crack) I'm quite emotional really! It's a thing that you don't really want to recognise it or talk about it and you think I know what I'll just carry on. Well you do but you don't. (Richard, 48, type 2.)

However a few participants were keen to remind people that regardless of the information that they have, and whether a diagnosis has been made or not, the Usher remains and it is up to the individual on how they chose to deal with it, "You can let it rule your life. And you can let it ruin your life. But this is the hand that you've been dealt with." (Dave, 46, type 3.)

6.1.4 **Additional medical conditions**

Participants in the research project were keen to highlight to others, that along with the Retinitis Pigmentosa which causes the vision loss in Usher, people with Usher are at a greater risk of cataracts, detached retinas, and cystoid macular oedema (CMO), creating their own problems and requiring treatment. Headaches, migraines, and fatigue were also commonly mentioned. Charles Bonnet syndrome is experienced as visual hallucinations as a result of the brain trying to piece together the images it receives as a person loses their sight. Rob explains it is important that people with Usher are told that they may potentially experience the frightening hallucinations as the brain seeks to fill in the missing gaps:

Regarding Charles Bonnet syndrome, when I was diagnosed I was already seeing things at the time. I thought I was going crazy! I thought that I was going mad! Not one doctor warned me that this is what happens, and I think that got to me the most, because I honestly thought that I was going crazy. But no one told me that this would happen. So if I'd known I wouldn't have... That was the main thing that I went into depression about. Because I was seeing things that I couldn't explain and I was also hearing things I couldn't explain. I'd walk out and I'd hear people talking and there's nobody around me... I thought I was going crazy. And no one told me, about this, and I

think that's what got to me the most. There needs to be more awareness on things like that. (Rob, 22, type 2.)

Furthermore individuals had a lot of unanswered questions about Usher, whether for example pregnancy, stress and ill health could hasten the speed of vision loss.

6.2 Types of Usher

Traditionally Usher syndrome has been organised into three types:

Usher type I (USH1) is the most severe form and is characterized by severe to profound congenital deafness, vestibular areflexia, and prepubertal onset of progressive RP. Type II (USH2) displays moderate to severe hearing loss, absence of vestibular dysfunction, and later onset of retinal degeneration. Type III (USH3) shows progressive postlingual hearing loss, variable onset of RP, and variable vestibular response. (Millan *et al.*, 2011: 1)

Individuals who do not fit into these subtypes are said to have “atypical Usher syndrome” (Stabej *et al.*, 2012: 27). Through genetic sequencing, see for example the National Collaborative Usher Study (Sense, undated) we see that “Usher syndrome is a clinically and genetically heterogeneous disorder” (Millan *et al.*, 2011:5). Due to these variations in Usher, participants whose hearing and vision loss did not ‘fit’ with typical definitions of Usher often felt confused about their diagnosis, Rosie explains in response to the question “Do you know what type of Usher you have?:”

I think it's two, I'm not 100% certain but I think it's two I can never match up because I know types, because I've seen them on the Sense website but it's like nobody actually fits into those categories so I think it's two. (Rosie, 21, type 2.)

Jess (15, with an uncertain diagnosis, possibly type 3) and her mother also speak about the desire to know what type of Usher Jess has:

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Mother: It's great when it's cut and dry and they'll say you're an Usher type so and so, this genotype brilliant.

Jess: Oh you might be this, or you could be this.

Mother: Which is what Jess gets. They say oh yeah I definitely think that you're a type 2, and then your central vision loss was the same time as your hearing loss so yeah you could be a type 3... but we're not really sure yet we'll let you know. So you think shut up! Why did you even say it if you're going to change your mind?! Don't move the goalposts.

Jess: I'm just special! I think that I'm type 50! They haven't discovered it yet.

Jess and her mother went on to explain how this had led to distress for Jess, so she had had some support from Sense in order to discuss her feelings about the lack of definite diagnosis. Five out of the 44 participants, had received a diagnosis of another type of Usher since their original diagnosis.

Four participants had been part of the National Collaborative Usher Study, as well as enjoying the opportunity to meet other people and take part in research they also found it 'satisfying' to know their exact type of Usher and where the mutation was found. One of the participants with Usher type 3 to some extent took pleasure in rarity of his diagnosis.

However some participants were not interested in the types of Usher as they felt that it did not affect their current situation. For example in response to the question "Do you know which type of Usher you have?" Richard (48, type 2) responded, "2. I guess it's not really important right now. Maybe further down the line when it becomes more important, I'll have a look at doing that."

There was some talk from a small number of participants about the desire to know exact gene mutations in order that when they are made available, participants will be able to take part in gene therapy. However participants were upset about the length of time that genetic sequencing takes, the associated costs, as well as the reluctance of medical professionals, as Nicola explains:

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I know a few Ushers that have had the blood test and it's been sent off to Germany to see if they can find the gene that causes Ushers, but me apparently, what they've said to me, is there's no point doing it because there's no cure yet. Whereas my dad's argument is well if you have it, you've got it, when there is a cure you can go I'm that. You know rather than waiting 8, 9 months or something, for results. Just that type of stuff annoys me, they seem to be reluctant to do what they can basically cos there's no point, because there's no cure, yet. (Nicola, 23, type 2.)

6.3 Living with the Diagnosis of Usher

After the shock of an initial diagnosis of Usher, accompanied by denial, depression and feelings of hopelessness, a lot of participants spoke about the need to get on with their lives and to live with a diagnosis of Usher. Crucial to carrying on with their lives was accepting Usher, and then moving forward to make adjustments and changes in order to assist them to live with Usher.

6.3.1 Accepting Usher

A characteristic of the participants in the research project was a desire to share their knowledge and personal experiences of Usher in order to reassure others and share good advice, this was particularly so with regard to accepting Usher. Michael, Alex and Kat commented:

Don't pretend that you ain't got Usher. Don't deny it. Don't try and be normal. I think it gets a lot easier once you let people know. Because you're only making it harder for yourself in the long run. I should know. I've been through it all. I think if you deny yourself that you've got the condition, you're only going to make it harder. And it doesn't make it any difference anyway, your life is still the same. I'm still a pain in the ass!! Before and after! (Michael, 34, type 2.)

You can't ignore it, I think the worst thing to try and do is ignore it completely. You can't carry on and think I'm alright as I am, you've got to try and accept it. (Alex, 29, type 3.)

I would say life just goes on the way it is. You've just got to move on. Act like you're normal. Everything's going to be ok. (Kat, 17, type 1.)

Accepting a diagnosis of Usher is not easy as Dave (46, type 3) explained, "The hardest thing ever is accepting it, but once you can accept it you can get on with your life. And you've got to get on..." Vicky (44, type 2) also reminds us that although a person may have 'accepted' that they have Usher it is still a very emotional issue, "if you ask me now it still makes me have tears in my eyes. I suppose, I dunno, it's been a rollercoaster, basically." Brian (40, type 2) reminds us that everybody's responses are different, "It's not a quick thing to understand. It's a very slow thing because everyone has different experiences and every single person is different from others because you won't get two the same." Furthermore not all participants had accepted their Usher, some were still reeling from the shock and hurting quite deeply, as the following quotation from Steven shows:

I feel quite bad. I feel quite bad... It's really, really difficult. Really really difficult. Cos the thing is because what I had before, and what I've got now is crap really. The quality of life... Obviously I'm rich in terms of children. I've got a fantastic wife. I've got my mum and dad and they're great. And my brother's fine. It's just that outside of that my quality of life is pretty rubbish. Things I know I would be doing if I didn't have this. Driving around. Taking the kids out. All three of them. Give my wife a break. It's difficult really because I can walk somewhere with them, but I'm not so comfortable especially with the 2 and half year old running around. Or he's pushing his scooter. Maybe he's running to go into the road. I'm not so confident with him. It hits your confidence. And you go into buildings, and you've got the kids with you, you've got the issue of it darker inside the building than it is outside. Then it takes you a while to adjust... There's no positives to it. I like to think that there is a cure. But I don't think that there will ever be one. Because there's not many people like me about. It's like what's the point in sorting me out. It doesn't make commercial sense. Sorry I know it's depressing... (Steven, 36, type 2.)

Participants framed their acceptance of Usher around different discourses. A common theme was to focus on the present as Chris and Olivia explained:

Who knows what the future holds. You could get run over by the bus tomorrow, before I even get to the stage where I lose my sight! You don't know what the future holds – no one does! You have to make the most of it. (Chris, 33, type 3.)

I just enjoy life, but maybe when I'm 80 or 90 I may go blind – I've got that at the back of my mind. But I'm enjoying my life now. When the time comes I'm going to have to sort that one out. **(I)** (Olivia, 54, type 1.)

Another strategy was to compare oneself to others. Participants were thankful that Usher is not life threatening, and recognised that other people are in more difficult situations, as Jess (15, type 3) and her mother explained with regard to the other children at Jess's school who also have serious medical conditions:

Mother: It's been actually quite good for Jess because you feel quite lucky that you've only got Usher.

Jess: I've got nothing wrong with me.

Mother: There's nothing wrong with her. We sit here and say oh gosh we're so lucky. Because so many of them are very very poorly. One of your little friends at school, she's very poorly with her condition, she was one of the first blind children that we met. It's all these experiences that makes Jess feel quite normal and confident with her sight loss.

Although not often spoken about directly in many of the interviews it must also be remembered that Usher is not the only thing happening in people's lives, which they also be in the process of accepting. Issues that were touched upon during the course of the interviews included sexuality, infertility, cancer, mental illness, and relationships with significant others.

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A third discourse that participants used to help them negotiate an acceptance of Usher was to look at the advantages Usher had brought them as Rob and Lucy explained:

I wouldn't change my sight now. I know that they've brought out the retina chip, and the gene therapy, if they offered me it, I'd say no. It's made me who I am today. It's made me a lot stronger person now, than I was before. I think I do marvellous. And I wouldn't change the way it is. I often get into debates with other blind people, where they say this chip will allow you to drive, to distinguish colours, see things. No! This is what I'm used to! If somebody turned round to me and cured my eyesight and I could see one day I don't think I'd cope... I don't think that I could go through that. But I cope now better than I did before. I'm a lot stronger person. I still have my down days where I think god you know I can't do this. But I just think, no I wouldn't change. Ever! I just wouldn't. (Rob, 22, type 2.)

I would never have become an audiologist. I didn't think that I could be, because I thought that I had to hear the sounds – which of course you don't. I'm good. It's not big headed but I'm good at my job. I have a much greater understanding than other professionals. (Lucy, 27, type 2.)

Finally the majority of participants were keen to point out that although they accepted Usher, in that they knew that they had it, they were determined that they were not going to accept its limitations. This determination to make the most from their situation is evidenced throughout this report. We also see that acceptance is an on-going process, and that as time passes and new situations arise individuals may have to go through a process of accepting their Usher on a new level, for example deciding to use a mobility aid, starting to learn braille, telling a partner about Usher. Ultimately however it is not a choice whether or not to accept that one has Usher, rather the way in which one chooses to live with it, as Hasan (15, type 1) explains, "Well I don't have a choice really. Well I've still got to be positive to continue as normal and to take each day day by day."

6.3.2 Adjustments and doing things differently

Making adjustments and doing things differently is a central feature of living with Usher, as Nicola (23, type 2) explains, "...it does get better, you've just got to adjust and find a different way of doing things." It can often be a light bulb moment when individuals recognise the need for change:

[Doctor] said use a cane. The Red Sea opened. I said well hang on a minute, I don't want to fate myself by using the cane. He said if you use a cane you don't need to say that you're sorry, people see you with the cane, they'll get out of the way, from their point of view and perspective it gave me a bigger picture. Then I came back to London, I see your point, this is what I need to do. I went back to my social worker and said I want mobility training, I need a cane. It's not a question of want, I need to do it, I need to prepare myself. I need to learn. I need to have adaptations. (Hamid, 44, type 2.)

Participants may go through a lot of anguish in building up to make a change, or slowly make small changes over a period of time, for example Carolyn (39, type 2) spoke about starting to use her cane whilst on holiday, then with close friends, before then using it in her local neighbourhood. Furthermore adjustments are often not made by the person with Usher acting alone and often incorporate friends and relatives, and professionals such as teachers, social workers, and mobility officers.

Participants took pride in thinking outside the box and being creative in making adjustments to living with Usher. Bethany (17, type 2) for example would take a photo of her bank statement and then use the enlarge function on her iPad in order to read her statement. Sally (42, type 2) bought a camping lamp that she used to take into restaurants so that she could see what she was eating.

Problems may also arise when there is not enough time, money or other resources to make adjustments. For example Steven (36, type 2) was thinking about learning braille, however with 3 small children he did not have the time available to learn. Doreen (53, type 1) wanted to go out with a communicator guide but could not find one who matched her interests.

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It can be difficult when doing something differently is obviously very different to how people usually do things as Jess (15, type 3) and her mother explain in relation to using refreshable braille display:

Mother: Jess could hook her Braille display up to her phone, but again it's one of those things that's very hard to sit here doing it different to everyone else.

Jess: I look normal when I've got my head phones on. And use my phone. But sit there using a Braille display!

Mother: I think as a teenager that is one of the most difficult things.

Jess: Maybe when I'm 30.

Mother: All the other teenagers can use their phone visually, or if they're blind by just listening to it. But Jess can't effectively. So she would then have to have her phone, her ear hooks, and Braille strip. It's just like you have to carry all of that around with you. And I think when you're a teenager you just don't want to. Do you?

Jess: No.

However, often there comes a time when doing things differently becomes the only option.

6.4 Discussion

This chapter has looked at the diagnosis of people with Usher syndrome. We see that when the participants were diagnosed with Usher, shock and denial were initial responses; this was especially so for the participants who were diagnosed during adulthood. The need for information, after diagnosis and on-going as individuals' experiences of Usher changes, was identified as particularly important. Participants spoke about the additional medical conditions that can be associated with Usher. The importance for some people to know their Usher type was identified, as well as raising awareness of the fact that the traditional textbook definitions of Usher types may no longer hold true. We then moved forward to look at living with a diagnosis of Usher, suggesting that acceptance of Usher is an important factor in doing so. This is not to say that all participants accepted their Usher, and acceptance of Usher was not a passive event. Furthermore participants would often pass through phases of acceptance as different hurdles

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were put before them. Nevertheless there was a strong desire by participants to encourage others with Usher to accept that they have Usher. Finally it was suggested that learning to make adjustments and doing things differently, although this may be difficult and the changes may only take place in incremental stages, is central to living with Usher.

Having explored the topic of diagnosis it is possible to identify three themes which encapsulate the lives and experiences of people with Usher. These three themes are:

- Change
- Predictability/uncertainty
- Diversity

These three themes will be used throughout the project to provide a lens through which we can further understand the lives and experiences of people with Usher. In relation to the topic of diagnosis we can see how these three themes can help to provide a framework to understand diagnosis and its impact of the lives of people with Usher.

6.4.1 Change

The point of diagnosis is, to the outsider, a moment of change. But this was not the case for all the participants. Those young people who had known about Usher for some time, or who had known about an eye condition, did not have a moment of change which might be called “diagnosis” (Daniel for instance took the discussion of Usher in his stride).

For others, mostly older people, diagnosis was of course a psychological change, but it was not an immediate change in how they lived their lives. Unlike a diagnosis of cancer or diabetes for example, there was no suddenly different lifestyle, no obvious change. Of course it may have changed their mental state, but there were no ‘physical effects’. In fact, change came later, as part of adjustment to actual changes in their vision, and the need for a cane, or an end to driving.

What in fact changed was their knowledge that things would change: the unpredictability of Usher. The adults who indicated that they might not have wanted to know, as a child, about Usher, are perhaps expressing their wish not to change their psychological state, their identity and security in who they are. In this they show the most fundamental change in diagnosis, that of ontological security, trust, continuity, predictability (cf Danermark and Moller 2008). Without knowing, they would not have had to change their world view and their view of themselves within it. Hasan, a much younger participant, perhaps expresses this also when he says that 15 year olds don't want to look at the future.

While the information from this group of participants cannot be seen as wholly representative, there appears to be a change in diagnostic practice through the years (as shown by the older and younger participants). This was also found in Oleson and Jansbøl (2005). Of course, since our research looked only at young people who **knew they had Usher** we cannot know what proportion of young people do indeed know. But changes in diagnosis do possibly mean that young people now know about Usher earlier than previously. The difficulties experienced through inadequate interpretation are perhaps less; Farah did feel that she knew what had been said.

6.4.2 Predictability/Uncertainty

As above, what had primarily changed for most people at this point was not anything in their lives, but their knowledge that things would change; the unpredictability of Usher. This is shown for example in the experience of Nicola, who discussed the ending of the relationship with her husband, because she did not know what was in the future. For others this was shown in depression and anxiety. In our sample the high levels of depression in other studies (e.g. Miner 1995 and 1997) was not so evident. Only 7 of the 30 adults said they had been depressed (although they were not directly asked this).

In most cases they managed this depression, sometimes with medication. While depression and anxiety are perhaps an expected outcome, they are not the overwhelmingly distinct feature of Usher. Depression may have been present, but it was overtaken by adjustment.

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For a number however, this was because they dealt with unpredictability by living in the present, not wanting to think about what would happen. This was shown by Hasan (as a young person) and also Kat and her mother, and Richard. As Kat's mother said 'we're off and at'em!'.

The unpredictability of Usher was certainly not apparently recognised by many of the medical staff who told people about Usher – the prediction 'You'll go blind', as they remember it being said, is not reflected in what has happened to the participants and does not reflect the range of experiences of vision loss which they then had. Chloe and Leah's doctor, while not giving them what they wanted to know – a timeline and outcome – was in fact more accurate.

Unpredictability, as already stated, is core to Usher. A number of people reflected that they were told the wrong information at or shortly after diagnosis, that it was too early for example, to know about talking books (Lucy) or canes (Nicola). However, the huge range of speeds of progression and the difficulty of matching type to individual and what this might mean for prognosis, means that a professional may not know what is useful to tell a person now.

One feature of the interviews was that for some, Usher did at least give one certainty – that it was Usher, and not something worse, which caused their vision loss. Jess too found strength in the fact that the only problem she had was Usher.

Particularly amongst the older adults, it seems that doctors told them quite blandly that they would go blind. While this is a firm prediction, in fact, this was not their lived experience, nor is it the case. For some, blindness came sooner than expected (Jess, Bethany) while others maintained a lifestyle which did not acknowledge visual impairment despite difficulties with vision (Megan). Even knowledge of the type of Usher did not provide helpful information about prognosis, as has already been noted, the previous typologies as they related to when someone might lose their vision did not apply universally at all. In addition, these diagnoses changed for a number of participants during the course of their lives (Though presumably, once subject to genetic testing, this is less likely to happen).

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Finally, in relation to unpredictability, it was not the case only that the progress of Usher was unpredictable. It sometimes influenced people so that they acted in unpredictable ways – getting married really quickly, or having a child (Nicola, Lucy).

6.4.3 Diversity

Diversity in the responses to this diagnosis was certainly present. Some responded to Usher with understandable depression. Some people have responded by embracing Usher, as a way of being, and understanding more about it. This includes those who say it has made them stronger, better people, and that they would not choose to be without Usher even if a cure was available.

For others, they have chosen instead to live in the moment, and not to worry about the future. Both Olivia and Kat chose to ‘act normal’ and then deal with issues later. Some made decisions to do certain things – such as see the world, get married, have a baby, or to raise money for causes related to deafblindness – either earlier than they would have or perhaps things they would not have done at all, but for Usher.

7 Mobility

Mobility is an important aspect of everyday life, and for people who are deafblind their hearing and visual impairments can have a significant impact on their mobility (Parker, 2009). Mobility, or orientation and mobility, is the ability to travel safely and effectively as a visually impaired person. Within this context, it includes specialist techniques such as using a long cane, with strategies and contexts, such as room orientation, using public transport and using a guide dog. Orientation and mobility applies to both indoor and outdoor skills, the knowledge of how to move safely and efficiently around their home (for example to find things) as well as to travel long distances e.g. on a bus.

The importance of mobility and the challenges that it brings to people with Usher can be seen in the strength of feeling from this quotation from Victor:

Overhanging branches is a real issue. Walking in the street with a cane and walking into thorns and branches hanging down. Cyclists on the pavement when I'm walking with the cane, left to right. Cyclist comes along and saws off a third of my stick. Straight through it. You can't walk the streets, broken pavements, overhanging branches, A-boards, cyclists. That's one of the reasons that I rely on door to door transport. It's hopeless! Absolutely hopeless! They won't enforce it, and I know why, because it's a vote loser! They'll lose votes and people don't want to pay. I've told the council loads of times to help me and they don't. Lip service. Mobility is an issue. Putting one foot in front of the other I can do that. They tick the appropriate box. But that's ridiculous! That's why I'm screaming at these stupid people at Atos and the government. They have to use more taxis, and if I can't get dial-a-ride I need to get minicabs. It's all extra money. Going to the shops sometimes, I'm tired. I can't face it. Even a short walk to the shops with overhanging branches I have to get a minicab. All of these things have to be paid for. (Victor, 56, type 3.)

Participants had lots to say about mobility. We shall begin by exploring mobility in the home, followed by at school, then with regards to public transport. We shall explore participants' thoughts and opinions on using

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and not using mobility aids such as canes and guide dogs, before considering driving and its significance for people with Usher syndrome. The majority of participants spoke about mobility in terms of vision; however we also explore the effect of deafness on mobility. Lastly we shall examine the specific nature of Usher syndrome and its effects on balance, and consequently its impact on mobility. As we cover these topics the three reoccurring themes of change, predictability/uncertainty, and diversity will be explored in order to provide an insight into the lives of people with Usher.

7.1 Mobility at home

Generally within participants' own homes mobility was not an issue; they remarked that they could get around easily, but nevertheless adopted strategies to keep things tidy and to know where things were. As Sally (42, type 2) commented, "Familiarity is half the battle... You know where everything is. You're so precise. You've got a place for everything. You train yourself."

Other members of family, or visiting friends however often needed to be reminded to keep obstacles out of the way or put things back in the same spot. As Carolyn (39, type 2) explained, "My daughter does karate and my son does swimming. When they come in they make sure all their stuff is upstairs not in my way. If it's on the floor I'll trip over."

Going to other people's houses was recognised as more difficult as Michael commented:

I like my independence, and I guess that it all goes with Usher, but if someone asked me round to dinner, I wouldn't really want to go because, I don't really know their house. I don't know how their kitchen is. I'm scared of eating in front of them in case I lose a pea or something and never see it again. I hate that, it makes me feel... I feel self-aware and I tend to avoid. That's why I don't really like going round people's houses and stuff like that really. Unless I really, really know them. And they don't mind me bashing into things. (Michael, 34, type 2.)

A number of participants bypass these difficulties by using *their* homes as a base for their friends to gather, or only going to the homes of very close friends. Participants were often very self-conscious of their mobility difficulties, even to the extent that they avoided taking part in activities as it would expose them, and their difficulties, to others, as Chris explains:

You can't on a date, after knowing them for two hours say, can you take me to the bathroom please?! ...The thought of getting up in a pub that is totally dark, and somehow hope that you end up where the toilets are. There are times when you chose not to do something when you are on your own – definitely! (Chris, 33, type 3.)

7.1.1 Lighting

The importance of good lighting was mentioned by a number of participants, in particular the difficulties associated with energy saving light bulbs and the cost of electricity in keeping lights on all day as Olivia explained:

I pay £900 a year for electricity. That's a lot of money. It has to be bright. And everything's bright. I can't see if it's dark. I've been trying to save electricity, and I do believe for environmental reasons we should be doing that. These lights in here are halogen, they're bright, they're suitable, but they're really expensive, the led are really cheap but I can't see anyone. So I'm stuck in the middle really. **(I)** (Olivia, 54, type 1.)

The mother of Chloe and Leah also commented:

Ah the electricity board must love us, first thing they do when they come through that door is put the lights on. I think if they can do it for telly, they can do it for lights. Do a reduction. (Mother of Chloe and Leah, 16 and 19, type 2).

The issues around energy saving light bulbs, and the lack of understanding/support from the Government were also brought up by Dave (46, type 3) and Sally (42, type 2) in their joint interview about their experiences of Usher syndrome:

Sally: 100 watt bulbs everywhere.

Dave: I bought the last ones!

Sally: I did! I've got a cupboard actually, with a box full of them.

Dave: These idiots that decided 100 watt bulbs were no longer going to exist.

Sally: That's awful. The Government should never have gotten away with that. It's like banning wheelchair ramps. It's as severe as that for some people with sight problems. Awful. I don't know how they've managed to get away with that.

A number of participants' homes had been assessed by local sensory teams, and appropriate lighting had been installed as Rebecca commented:

My flat has special lights, when I moved it didn't have any in, I had to ask the housing association to help me change all the lights, made them much brighter, so easier access and I could see much better then. **(I)** (Rebecca, 29, type 1).

Harry's (15, type 2) mother was particularly impressed with the new en suite bathroom that had been installed in Harry's bedroom, complete with sun tubes to provide natural light, and extra blinds on the windows, made possible through funding from the local authority. Although a number of participants had successfully managed to obtain the appropriate lighting that they required, not just bright lighting, but natural lighting, and the ability to dim lights as necessary, some participants were still struggling with assessments. Getting the appropriate support was a challenge as Bethany's mother explained:

...I thought it was about time that social services got involved, did the assessment they should have done six years ago, and you know, made available things that can be got for her for free. That are sort of over and above what we would do as a family, and so far they have done bugger all! (Mother of Bethany, 17, type 2.)

As well as better lighting, Bethany's mother was hoping to get funding in order to repaint the house as the current paint colour was not suitable, however deciding on a colour was not a straight forward task as Bethany's mother remarked, "the [paint] that I really liked she was like 'I couldn't cope with that. It's just too bright.'" And I thought grrr! Usher

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syndrome takes over everything! So we had to choose a colour that she could deal with.” Such a comment serves to highlight the way in which family and friends often have to make changes and adjustments because of an individual’s Usher, not just the person with Usher.

7.2 Mobility at school

Mobility in and around school, as well as lessons from mobility officers, were specifically mentioned by the young people who were still in school/college. For the adults involved in the project, mobility in school was mentioned in passing, as will be explained in more detail later, this was often in relation to being clumsy and being teased for not being able to see as well as other people. The experiences spoken about by the young people in the project varied from excellent to pretty poor, further highlighting the disparities in experience for people with Usher syndrome. Some solutions to improving mobility were pretty straightforward as Hasan noted,

My school is quite an historical building, it has quite a lot of history, so there’s a big tunnel which is quite dark. It’s a big tunnel between classes. But that’s solved now because apparently in the summer, they’re putting lots of lights in there now. (Hasan, 15, type 1.)

For Ryan (17, type 2), the introduction of lights, and yellow strips on the stairs was well received, “It was all dark. But as soon as I started college it all changed. Yellow tape on the floor. Lights in the dark corridors. So it was actually really cool.” This was important to Ryan as he disclosed that before he started at college, “[They] had this darkness bit. And I was getting all worried. But they actually put corner lights in before I went to college. And it helps now.” Ryan along with Callum (14, type 1) and Jess (15, type 3) (who all took part in further discussions on mobility in a second interview), also mentioned not wanting to be singled out for their mobility needs with regards to bags being left on the floor. All three young people spoke about their experiences of bags being left on the floor, and tripping over, or fears of tripping over. They remarked that it was important for teachers to remind other students to hang up, or put bags away properly, in a manner that did not single out individuals but

as a more general health and safety policy within the school/college.
Jess and her mother expressed their feelings:

Mother: And do you think you'd like it if everybody got told, 'pick your bags up Jess is coming through!'

Jess: Yes and no.

Mother: Why?

Jess: Cos then I'm not going to fall over them.

Mother: But why no?

Jess: Because I'm normal. Don't want to ask for the bags to be moved!!

There were some examples of good practice from within schools, for instance Callum's mother reported that Callum had been having some difficulties in PE which his class teacher noticed, and then contacted the sensory impairment team who arranged for an assessment of Callum's needs at school.

Farah (17, type 1) also spoke of the excellent support that she received from both the person providing her mobility training and the interpreter in order for to learn how to use her long cane, and to build up the confidence to use her cane out in public:

I didn't like people looking at me. But the teacher said to try. And I try. And I did I got used to it. (mobility worker) and an interpreter came to my home, and I was a bit fearful at first, I didn't have any confidence, I didn't like people looking at me, but as time went I got comfortable with it. I used it when I took my exams. **(I)** (Farah, 17, type 1).

However as Farah goes on to comment and will be discussed later in the section on using a cane, on-going training is important as people's needs change, as well as the fact that mobility aids are not a cure-all for an individual's mobility needs,

I need more training, I'm not actually 100% confident with (my cane). I need more training. At night I don't. It's a risk. I don't really go out at night. **(I)**

Despite the generally positive comments on mobility in school a couple of participants, both of whom had significant vision loss reported

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negative experiences in relation to mobility whilst at school. One participant (a young person) reported that access to a guide dog was denied at critical points and for extended parts of the day which cumulated in this participant leaving the school in the middle of A-levels. This participant with the family described a lack of understanding by the school, which was a residential school for deaf children, on the needs of pupils with visual impairment. The participant commented:

I know that [the school] really are in the wrong for doing what they did cos. I don't know why, they were just so biased towards this [pupil]. They were so worried about his safety, you don't care about mine, and that's what got to me. That's all what the SENCo said to me one day, you know keep an eye out for [pupil with allergy]. 'Who's the blind one?' He sort of went 'huh'. And it obviously sunk in and he thought, yeah I shouldn't have said that. I was like yeah, who are you trying to protect here.

The consequences of being denied the guide dog, and then having to leave school were very challenging for the participant :

I left in the November time. It was horrible. I just felt as though I wasn't part of the school any more. It really, really affected me. Like I'm going counselling weekly because of it. It kind of affected this whole other... I've got this major thing about self image now. Like I don't... That's why I struggle leaving the house with [guide dog] because I feel as if everyone's going to judge me because I've got this guide dog. Cos being at [school] it's like you've got a guide dog you've got to be in be put in this isolation room... It's taken a long time because I was blaming [guide dog], taking it out on [guide dog], and I was saying oh I want her sent back. Cos I just thought it was the easiest solution. Not because of my safety or anything. I'd be putting my safety at risk, if I sent her back, but I was just like if I send her back it'll make life easier. But I stuck with it.

Jess (15, type 3) who attended a residential school for deaf children "fell over because it was dark and I couldn't see" breaking her wrist in the process, demonstrated the very real consequences of poor lighting and not using mobility aids when necessary. What made this more difficult for Jess' mother was the lack of understanding and awareness of issues

relating to mobility for students with Usher, despite the school being a residential school for deaf children:

[School SENCo] said oh we can only apologise. No I don't want you to apologise – do it! Do something about it. What are you going to do? It's danger! Encourage Jess to use her cane. She's got a cane...
...And do you know she never got that cane out once whilst she was at (school). Because she hated it while she was there. Because there was so much lack of blind awareness, Usher awareness. So much bullying and cliqueness... And you'd rather break your neck than use your cane. The amount of times that she fell over. (Mother of Jess, 15, type 3).

7.3 Mobility and public transport

Mobility in relation to public transport was another area that was commented on specifically by participants in the research project. For participants the issues relating to public transport were twofold, firstly the availability, or lack thereof, of public transport, and secondly the issues relating to access on the transport, for example catching the correct bus or train, reading timetables, and boarding buses.

Generally for participants living in urban areas public transport was not seen as a problem, whereas for those living in more rural areas, or towns less well served by public transport there were difficulties. Cohen (2009), writing about Rebecca Alexander, (who has Usher 3), in the New York Times said “The young [blind] flock here for the opportunity to play on a level field, where everybody is dependent on public transportation, cabs, deliveries, and Internet ordering”. Richard, living in a large city commented:

I'm registered partially sighted with the (local) association for the blind... I get a Freedom Pass which means that all tubes, trains and buses are free for me to travel.... It's a bit of a pain sometimes going to some places. But I can get around. (Richard, 48, type 2.)

Megan, also living in a large city mentioned the ease with which she could use public transport:

I use the underground mostly. So convenient. Nobody has a car in London; nobody in my age group does. Everyone gets buses, taxis, underground. It's SO easy. So I don't feel like I'm missing out.
(Megan, 24, type 2.)

The experiences of people living in less urban areas were less positive. For example Lucy (27, type 2) actually went as far as moving house from a village with poor public transport connections to an urban area with regular buses and a train station just across the road. Julie (38, type 2) tells of her difficulties accessing public transport in her local area, "The public transport is rubbish really... It's alright saying use public transport but it's just not any easier it makes your life a lot harder."

Even when public transport is available, people with Usher still face difficulties in accessing the transport, for example despite there being a good service in her area, and using public transport regularly Megan found it difficult to know which stop was hers, especially when the recorded announcements were switched off.

Dave's comments highlight the fact that although there might be a good bus service available, getting to the bus stop may represent a huge challenge:

It's like a hidden disability. I dread the dark nights, early mornings, if I get the bus home, the bus is 200 yards up the road, but me getting those 200 yards, you just want to crawl on your hands and knees. You just want to get there as safely as possible, without bumping into somebody, without having to apologise. (Dave, 46, type 3.)

Public transport services are obligated to provide additional support on request for passengers with disabilities, however when this support does not materialise, passengers can be left stranded, which has an impact on levels of independence, and means that individuals face higher costs, as Rob explains recounting his experience with local transport:

Transport is really quite difficult. Cos I can't get to places where I want to go. Cos I'm fairly independent and I like to do things on my own if I can. But things like trains and buses it's just so hard... I went to (large city) and I rang up and I booked in advance the time of the

bus that I wanted to leave and if they could just plonk me on the bus at the bus station. They said oh yeah, yeah, we'll do that. So I gets there. I missed my bus. Nobody turned up for me. And I ended up having to get a taxi home, so that's the last time I'm doing that! It's just things like that... (Rob, 22, type 2.)

The extra costs associated with transport were noted more frequently by those participants whose visual impairment was more advanced. Victor and Susan had been involved in campaigning to raise awareness of the particular mobility needs of deafblind people. Susan commented:

I went down to London to Downing street to talk to the MPs about my bus pass. I campaigned and challenged it because I was not able to travel before half nine in the morning. I was successful. They reinstated it and upped my pass to a 5 year one, not annual one. (I) (Susan, 48, type 1.)

Taxis in particular attracted comments from respondents. On the one hand, as spoken about by Vicky (44, type 2), Steven (36, type 2) and Alice (41, type 2) taxis were seen as a convenient means for getting from place to place. On the other hand there were disadvantages as Lucy (27, type 2), a guide dog owner, described her experience of discrimination from some taxi drivers, although she admitted that there was often another taxi driver around to 'force' a driver to take her and her dog "part of me just doesn't want to bother." Furthermore taxis were seen as an expensive extravagance, but as Vicky noted, due to the fact that she was no longer able to drive because of her Usher, if she wanted to take part in activities and to get home from work, taxis were often her only option. As Vicky had begun to accept her Usher more, and the changes that it required of her, the more confident and satisfied with the situation she had become, especially when she balanced the cost of running and maintaining a car with the cost of a taxi journey

...the way that I see it is that I don't spend a fortune on a car. Cars are expensive nowadays. Petrol. So I just think if I have to go somewhere, and my husband can't give me a lift or whatever, I just get a taxi, and I don't feel... because taxis are always seen as a bit of an extravagance, aren't they? But I don't see it as an extravagance. Because I'm not paying for a car. (Vicky, 44, type 2.)

Nevertheless Vicky too had encountered difficulties with taxi drivers, but saw it as her mission to educate others about RP and other visual impairments. In order to do this she used the same taxi company with the same driver and asked him to put the interior lights on so that she could make her way to the cab safely.

7.4 Mobility aids

Examining the mobility aids that participants used most regularly highlights the diversity of experience with regards to the lives of people with Usher. The most common approach/strategy was to have no mobility aid at all with 60% (25) of people not using any mobility aids. However when this data is examined more closely we see that the responses vary by age, 67% (8) of young people, 67% (10) of younger adults, and 47% (7) of older adults do not regularly use a mobility aid. Some participants did however have a cane (either long or symbol) that they rarely used or kept in their bag 'for emergencies'. When we look at the figures in relation to Usher type, 67% (10) of people with type 1 Usher did not use a mobility aid, 55% (12) of people with type 2 Usher did not use a mobility aid, and 60% (3) of people with type 3 Usher did not use a mobility aid.

Participants who preferred to get around in the day time without a mobility aid spoke of their difficulties at night, in dark places such as pubs, cinemas, and restaurants, and in unfamiliar areas. Linda spoke of the variables that affected her choice of whether to use a guide or not:

Yes if it's an area that's familiar to me and I know the area well. If the weather's ok, if the light's ok. If the light's not so good or it's dark I need to be guided. It varies. If I know the area well I'm happy to go out, but if it was London, absolutely no way I could do that on my own because I would need to be guided. Because it's so busy. **(I)** (Linda, 46, type 1.)

Or sometimes an individual needed some support at a particular moment – Faith (23, type 1) and Rebecca (29, type 2) spoke about wanting an arm from a friend when entering the cinema.

5 people (12%) had a guide dog. These were 2 young people, 1 younger adult, and 2 older adults. Four had Usher type 2 and one had Usher type 1. None of the guide dog users were BSL users. Two of the younger adults were on the waiting list for a guide dog and two of the older adults were waiting for an assessment to see if they could go on the waiting list for a guide dog.

People who had the most significant combined hearing and visual impairments were most likely to use a communicator guide. Overall 5 people (12%) had a communicator guide. Four were in the older adult age group, and one was in the young people group. Two of these had type 1 Usher, one had type 2, and two had type 3 Usher. Other participants also used the services of communicator guides, however for these 5 people a communicator guide was their preferred (or only) means of getting out and about. As will be discussed in section 7.6 on hearing impairment and mobility, a communicator guide is much more than a mobility aid, - the use of communicator guides demonstrates the complexities of combined hearing and vision impairments..

7.4.1 Using a cane

For the majority of participants using a cane was not a straightforward act of using a tool to enhance mobility. As Sally (44, type 2) and Lucy (27, type 2) noted a cane is the 'symbol of blind people' and tells others about the vision status of the person using it. This may be useful as shorthand in certain situations, for example when getting on a bus.

However due to the general public's misunderstandings of what blind means participants were anxious not to be seen as 'trying it on':

I think if I use a cane, people will be, oh I've just seen you texting on your phone, or reading a book. Why are getting off the train with a cane? Are you just trying to get an advantage over us? (Alex, 29, type 3.)

This is compounded by the fact that Usher is a hidden disability, as Jess's (15, type 3) mother put it, "She doesn't look blind." Although speaking from the perspective of a guide dog owner, Alice shared these opinions:

But like a lot of difficulties you look normal, people don't realise. I had an old man one day at the bus station, sidle up and say you don't look blind. So then I had to explain to him, but I still think he was none the wiser really. (Alice, 41, type 2.)

Also the notion of 'blind' did not fit well with participants who still had some vision. Hamid (44, type 2) for example spoke about not wanting to "fate myself" by using a cane, and according to Callum's mother (14, type 1) he had refused cane training because he could 'still see.' One participant also spoke of getting a cane because she thought that she had to in order to access Direct Payments.

The progressive nature of Usher also made it difficult for people with Usher to 'come out' as blind. Julie (38, type 2) had thought about getting a cane but was anxious about what her neighbours would think as one day without the cane she could 'see', but the next day with a cane she would be 'blind'. Nevertheless as Carolyn (39, type 2) comments there often comes a point when using a cane becomes necessary:

I went to the supermarket and I turned round one day and I didn't see the trolley, this man just turned round and shouted at me in the middle of Morrisons "can't you see where you're going." I tried to get out of Morrisons and that's when I realised that I needed to use something to help me to let people know I'm having problems with my sight. (Carolyn, 39, type 2.)

Carolyn, like Dave (46, type 3) however, took a step by step approach to starting to use her cane. They spoke about how using their cane for the

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first time away from home whilst on holiday, in order to get used to it before facing people that they knew.

Sally (42, type 2) and Dave (46, type 3) also spoke about coloured canes. They saw them in a positive light, more as an accessory than an aid. Sally commented, “You can get like a pink one and match it with your outfit.” Here perhaps because individuals were taking ownership of their cane, albeit in a small way choosing its colour, they were happier to use one.

Although participants were not asked about it specifically, a few participants spoke about the use of red and white canes to indicate that someone is deafblind. Nicola (23, type 2) had opted against a red and white cane as she felt that no-one understands what it means. Likewise Alice (41, type 2) had opted against a red and white harness for her guide dog, as she felt that it would not add any additional information. Additionally Steven (36, type 2) as a deafblind person felt that other people had a lack of awareness of what a red and white cane signifies, by talking about red “stickers” and he spoke of wanting to take them off when he got a free moment.

A number of participants were reluctant to use a cane as they felt that it would signal to others their vulnerability. Olivia commented,

I don't mind using my cane when there's lots of people around me, but when I'm on my own I don't feel safe because you're open to being robbed. It happens so many times. **(I)** (Olivia, 56, type 1.)

Likewise Megan (24, type 2) used a long umbrella as a cane in order to avoid looking vulnerable. Olivia was also reluctant to use a cane, since as a Deaf person and BSL user she wanted this to be her primary identity, rather than being blind. She was concerned that people would immediately think that she was hearing and start talking to her, if they saw her with a cane.

Learning to use a long cane correctly takes time, dedication and a lot of concentration as Nicola (23, type 2) and Farah (17, type 1) observed. There is also a need to ensure that cane training suits an individual's particular needs. Rob (22, type 2) for example found that the cane skills that he had been taught were not applicable to his rural environment.

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Nicola also highlighted the importance of night time training. She had had her training in the summer “when it doesn’t get dark til 10pm” consequently, “You might have seen the shop (about 250m) away, I don’t go there on my own in the dark. I’m a hermit when it comes to dark.”

Even when a person uses a cane on a regular basis it still requires effort and concentration, often leaving an individual with fatigue as Steven explains:

Another thing is that it does take up a lot of your mind. More often than not I’m always thinking about I mustn’t walk into this, I’ve got the stair coming up here, I’ve got the table here, which I need to make sure that I walk around, and make sure that I don’t walk into anybody. That takes up a lot of what goes on in my mind. Then of course I forget about the little things. Don’t forget to do this, don’t forget to do that. So it is very, very, very stressful. You always get caught out. It’s one of those things really. (Steven, 36, type 2.)

Lastly, the personal attitudes of those teaching mobility was important, with both Ryan (17, type 2) and Daniel (15, type 2) commenting on how ‘nice’ their mobility workers were. Harry (15, type 2) and his mother were however frustrated with the lack of understanding shown by Harry’s mobility worker, as Harry’s mother explains:

Shouting at him because he had marks on his cane. Well when you bang things they are going to get marked! Again it’s not the understanding of how life actually is for them, you know. (Mother of Harry, 15, type 2.)

Sally (42, type 2) and Dave (46, type 3) also spoke of the insensitivity of rehabilitation workers, suggesting that they would have preferred to have had advice and training from another visually impaired person. As Sally said, someone who “understands what we are going through” (Sally, 42, type 2).

7.4.2 Guide dogs

Although only five of the participants had guide dogs, the majority of participants spoke about guide dogs and said that they were something to think about for the future. In fact Richard’s (48, type 2) family had

started puppy walking in order to get the children used to potentially having a guide dog in the future. In general guide dogs were seen in a positive light, and as preferable to using a cane as Nicola (23, type 2) and Abigail commented:

[People with Usher] are going out still. Pubs. Thanks to the dog. You know they can go out at night, you know meet. If someone asked me out for a drink, I'd be 'no' because I don't want to be the tag on, when I take my cane. Because a dog is a bit more sociable isn't it.

Everyone likes a dog. You know you can get out again. (Nicola, 23, type 2).

I think having a guide dog would be less embarrassing than having a cane I think. When I was 16 I had somebody to teach me how to use a cane, and I felt like everyone was looking at me. Yeah I think it would be less of an embarrassment to have a guide dog. **(I)** (Abigail, 29, type 1.)

Although a guide dog is a mobility aid it is still a dog and consequently might not suit everyone. Hamid (44, type 2) was disappointed that he could not get a dog because of his allergies. Farah (17, type 1) and Jess (15, type 3) were simply not keen on dogs. Steven (36, type 2), a father of 3 young children, did not want a dog whilst his children were small. A dog would not be suitable for Victor (56, type 3) as he had a cat and a small flat. Alice (41, type 2) also spoke of recent difficulties that she had been having due to the fact that her dog had been injured and was currently unable to work. Likewise when Alice had been injured she had to find someone to come in and walk her dog twice a day. Lucy (27, type 2) also commented that when she goes out with her husband and her dog comes too, she has to focus on working her dog, and cannot hold her husband's hand. For Vicky (44, type 2) who was in the focus group whilst Lucy was telling this story, this would be a very difficult aspect of owning a guide dog. A guide dog also has to be looked after and as Rob (22, type 2) explained, his guide dog had to be returned due to his depression and not being able to care for the dog:

I did have a guide dog but err... I was just diagnosed as blind. I was going through a very bad patch. I was battling depression, as well as

anxiety and stuff like that. Unfortunately due to that, I wasn't able to keep the dog. (Rob, 22, type 2.)

Alice also spoke of the challenges of becoming a guide dog owner and making this transition. Alice felt that there ought to be more support available for individuals and their families, as the following quotation shows:

There's been ups and downs. There's a lot to take in and such a big change not only to my lifestyle but to friends and family. I just sort of feel Guide Dogs should maybe involve friends and family a wee bit more. Like my best friend, she'd do anything for me, she found it a struggle, she used to help me a lot, but then all of a sudden it's me and (guide dog) and I can go shopping on my own. That bit I love, but it's hard for people around even when I go down to my mum's and that, she's used to helping me out more. I just say no mum, I can go off and do it. (Alice, 41, type 2.)

The young people in the project were particularly keen on guide dogs with Harry (15, type 2) commenting, "I want a guide dog but my mum won't let me. They're cool." However his mother's response was that he was not responsible enough yet. Dima (15, type 1) too was keen on the possibility of a guide dog however she knew that her parents would not like the idea due to the amount of work involved in caring for a dog. Ryan (17, type 2), perhaps like Dave and Sally who wanted coloured canes in order to have some ownership over their mobility aid, said that he wanted a guide dog, but only if it were a pug!

Guide dog owners also spoke of a lack of awareness on the rights and regulations surrounding guide dogs. As seen above Lucy, (27, type 2) and her guide dog faced difficulties when accessing taxis, and also in certain restaurants and Bethany (17, type 2) was denied access to her guide dog whilst at school. Of particular concern for Carolyn (39, type 2) and Alice (41, type 2) were attacks on guide dogs, especially since Carolyn's dog had been attacked previously. Carolyn reported however that the Guide Dogs charity had been supportive and had sent someone out to see her and her dog. In fact overall many participants were very positive about the work of the Guide Dogs organisation.

As shown above a number of participants were on the waiting list for guide dogs, and despite the fears of long waiting lists most people who had received a guide dog had done so reasonably quickly. No one had a dual trained dog (a guide dog also trained as a hearing dog for the deaf), however Rebecca (29, type 1) was on the waiting list for a dual trained dog. Lucy (27, type 2) had thought about having a dual dog however when she realised that the dog would be constantly working, and would not have that much opportunity to be a 'pet' she decided against it, especially since she could 'get by' with her hearing aids.

Lucy was an advocate of being a guide dog owner and in the focus group with Vicky was keen to answer all of Vicky's questions. Particularly of interest to Vicky was Lucy's experiences of bringing her guide dog to work and the fact that Lucy thought she may not be allowed to have a guide dog as she is 'only' registered as 'sight impaired' and still has reasonable amounts of functional vision. However as Lucy was quick to point out, ownership is based upon need rather than a specific degree of vision loss. By the end of the focus group Vicky was eager to start the application process to become a guide dog owner.

7.5 Driving

Driving and giving up driving one was of the most significant changes for the participants in the research project. The biggest challenge was the lack of independence and the reliance on others:

I'd like to drive! That's my main bugbear. I'd love to be able to drive. It'd be just nice to have a bit more freedom. I'm just finding it more and more, now I am starting to get worse, I just find it more and more restricting. And it's frustrating. My husband got a lift this morning and the car's sat there on the drive, and I can't use it! (Julie, 38, type 2.)

If I had good vision [driving is] the first thing that I would do! Because I hate relying on my mum for lifts – I want to be independent, but I have to rely on my mum for transport and lifts and things. **(I)** (Jayne, 28, type 1.)

Parents in particular found it difficult, because not only could they not take their children to places; it meant that their partner had to take on a

greater share of childcare. Participants (and their mothers) also spoke of learning to drive as being one of the markers of transition from childhood into adulthood, and for young people for whom this was not possible it was particularly difficult. Alex spoke about his experiences:

Learning to drive I say is probably the biggest thing if you say do you miss out on anything, I'd say that because there was a time obviously when I was losing my sight and all my friends were getting their driving licence for the first time, and it was such a big thing for them. I've got my freedom! I've got this! I can wherever I want! I can jump in the car and drive off to Wales if I want to, and I'm like ok I haven't got that, I don't know what that feels like. So I sort of feel as though I missed out on that. (Alex, 29, type 3.)

Bethany's (17, type 2) mother said the worst age is 17, "all her friends are learning to drive."

For Ben (30, type 1) and Erin (30, type 2) it was only when they wanted to learn to drive that the meaning of having Usher had any impact. Rosie (21, type 2), Chris (33, type 3), Alex (29, type 3), and Ben (30, type 1) spoke of never having driven and consequently suggested that they did not feel as much of a loss as other people with Usher who had had to give up driving. Ryan's (17, type 2) mother was also struggling over Ryan's desire to learn to drive – he had the provisional licence but would it be better for him never to learn. In the meantime, whilst coming to a decision, Ryan had been riding motorbikes off road.

In adapting to change or as a result of not being able to do what a great many people do, some participants chose to see the benefits of not driving, and expressed positive comments. For example brothers Chris (33, type 3) and Alex (29, type 3) spoke of being healthier and fitter than their driving friends, as well as having better knowledge of public transport systems. Other participants said that they liked not owning a car as it was better for the environment, that they could have a drink whilst on a night out and that it was cheaper as they did not have to pay for things such as petrol and car tax. When Alice (41, type 2) had to give up driving she sold her car and used the money to go on holiday saying, "Get some good out of it you know." Lucy (27, type 2) was keen to reinforce the benefits of subsidised travel, remarking having Usher "[is]

not the end of the world.” Michael spoke about the benefits of using public transport:

I don't miss it that much. Some days I do. When it's raining. But I quite enjoy walking around anyway. Keeps you fit don't it And I think you meet more people when you go out and about. In a car you just sit there. (Michael, 34, type 2.)

Hamid also spoke about consoling another person who had recently been diagnosed as having Usher:

...so I said look on the positive, you don't have to pay tax, you're saving money, you don't have to do that. “oh yeah, yeah you're right”. Think about road rage – you're not stressed out. Ha ha very funny. It gave her something to cheer up and she was happy with it. (Hamid, 44, type 2.)

Participants who had had to give up driving spoke of how difficult it was to surrender their driving licence, with Michael (34, type 2) and Dave (46, type 3) keeping theirs just in case a cure was found. No one spoke of carrying on driving after being advised not to, however a couple of participants did speak of near misses before being diagnosed, and difficulties with driving is what compelled Richard (48, type 2) to get his vision checked, which led to his subsequent diagnosis of Usher. Vicky (44, type 2) and Lucy (27, type 2) in their focus group spoke of the final letter from the DVLA on surrendering a licence:

Lucy: It was a horrible letter. There was no sympathy.

Vicky: I think that's a really hard thing to deal with. Knowing that at some point you will have to stop driving. And you're just hanging on to that. And when you finally get that letter you're like, that door's now closed kind of thing. Sorry makes me feel quite emotional thinking about it because it was such a big moment.

7.6 Hearing impairment and mobility

The majority of participants spoke about the challenges of mobility in relation to their visual impairment, however some participants spoke about how their hearing impairment also impacted upon their mobility.

Mobility

Olivia (56, type 1) and Barbara (50, type 1) both Deaf sign language users, had been thinking about getting a cochlear implant to help with environmental noise, in order to support their mobility:

The reason why I've been thinking about my cochlear implant is because to make sure that I am safe. To make sure that I can hear things coming. Or if I walk the dog I don't hear noises from behind. And sometimes I miss her, but if I had a cochlear I would be able to hear her footsteps. And roads. Crossing roads. Trains – sitting there and not being able to hear the announcements being made. I'm on edge waiting for the announcements, but I could just sit down and read. Sometimes someone comes and puts something in front of me and it makes me jump, and I say sorry I can't hear, and it's so embarrassing. **(I)** (Olivia, 56, type 1.)

Rebecca (29, type 1) also had a cochlear implant and Jayne (28, type 1) had recently had bilateral cochlear implants in order to give her access to environmental sounds, which they suggested made them feel safer.

Jess (15, type 3) and her mother also spoke of the difficulties with the most modern hearing aids which focus on speech, leaving out environmental noises which are crucial for a person with Usher, as Jess's mum explains:

...the new digital hearing aids, particularly for people with Usher – utter nightmare! Because they suppress background noises. And that is the worst thing in the world if you can't see. It takes away all your environmental clues. Whether it's traffic noises, kids running up the road, somebody shouting, whatever's going on around you.... It's crazy when you've got Usher! Silly! When are you going to know that it's safe to cross? (Mother of Jess, 15, type 3.)

Electric vehicles were also problematic for Victor (56, type 3) and made it difficult for him to access the outdoor environment, as he explained, "The silent vehicles is another issue – the cars that you can't hear. I can't see them. And now I won't be able to hear them. So what hope is that?" Bethany (17, type 2) spoke about how as a younger child she was not allowed to walk to school because could not hear the traffic.

Communicator guides were an important means of getting out and about for participants who had significant hearing and visual impairments. Communicator guides provide support for both mobility (guiding) and communication, as Susan who was a hands-on BSL user commented:

I used to use a cane before. Now it is easier to work with my communicator guide because that way we can have continuous contact so we can talk if we need to. **(I)** (Susan, 48, type 1.)

Communicator guides were seen by some as vital. Susan has a communicator guide for 53 hours a week. Without a communicator guide Susan is stuck indoors. Unfortunately her communicator guides do not work on Saturdays or Sundays, so Susan is left indoors unless her mother comes to visit. Brian (40, type 2) however preferred to use his guide slightly differently – he liked his guide to step back a bit allowing Brian to go about his business, but comfortable in the knowledge that support was available should he need it.

As communicator guides provide such a personal service it is important that they fit with the needs of the person they are supporting, however for Doreen (53, type 1) and Jess (15, type 3) this was not always the case. There was a personality clash between Doreen and her communicator guide which meant that she was left without support and consequently had to rely on her husband should she want to go out:

I had comm guide before. Stopped now. The woman who was my comm guide was very different to me. Our personalities didn't match. I like woman's things, she was a tom boy. She wasn't interested in going shopping, or looking around shops. There was a bit of a personality clash. We got on ok, but I wasn't very comfortable. **(I)** (Doreen, 53, type 1.)

It was important for Jess and her mother that Jess's communicator guides were young and female, but they also had to have knowledge of hands-on BSL as this is how Jess communicates in situations where she has hearing aids out, for example at the swimming pool or on a rollercoaster. Jess also had the added complexity of finding suitable communicator guides in her own area, and whilst at residential school.

7.7 Balance

Participants with type 1 Usher, and some participants with type 3 Usher, often have balance difficulties due to problems with the vestibular apparatus (Millan *et al.*, 2011.) Participants spoke of being mocked for the way that they walk, for being clumsy. Jayne (28, type 1) commented, “And my balance is terrible – I look like I’m drunk sometimes!” Hasan (15, type 1) spoke of being embarrassed at school because he tripped over easily. Because of Ryan’s (14, type 1) balance issues he had not been able to learn how to ride a bike. For Rebecca (29, type 1) it was her walking (and nightblindness) that led friends to discover her Usher before her parents had made her aware of it. When Jayne finally understood about her Usher and her difficulties with her balance, it took her back to experiences from her childhood:

What’s really important is that their family need to understand, because it was really difficult for me because my mum didn’t really explain things to me. As I’ve got older I’ve realised this. I don’t blame people, but I have realised this as part of life. What families really need to know, like I used trip on things and they’d say ‘watch where you are walking’ and that really was really annoying. That’s one of the problems. People say that because they don’t understand. **(I)** (Jayne, 28, type 1.)

Jess’s mother also spoke about Jess’s balance difficulties and explained how it contributed to Jess’s fatigue:

I think sometimes there’s just too much going on, to walk to talk, to keep your balance, keep yourself upright. It must be quite hard. And then still be expected to keep up with the world. At school, and at college and everything. (Mother of Jess, 15, type 3.)

Vicky (44, type 2) made the point that it is not always because of Usher (balance difficulties or visual impairment) that an individual needs support, “You need to check whether there was something else going on or just Ushers.”

7.8 Discussion

7.8.1 Change

Of all the issues which participants in the project examined, issues around mobility were one of the most emotional and those which induced the greatest change. Issues relating to mobility afforded participants with a reminder of a way of life that they once had, and acted as signifier of change in the future. While practical issues were of importance –how will I get to places - of even more significance were the uses of identity and ontological security. Changes in their vision and the subsequent effects on their mobility altered people's perception of who they were. Comments such as 'I'm normal' (Jess), indicating that she feared she was not, and 'everyone's going to judge me' (Bethany) because she had a guide dog) and Olivia, who felt she would somehow be less Deaf if she used a cane (and so was recognised as visually impaired). Difficulties in mobility changed who they were because of activities they could, or could not, get to. They might not go out to friends' houses, they might not take part in their leisure activities (see chapter 12) or they might not be able to get to work.

They were also changed by the perceptions of other people towards them. This changed their own concept of themselves. They might choose to use a cane but feel they had to justify it – because they were not 'really' blind – or to justify it to others, who might think they were not as visually impaired as they were making out. Using a cane might make them feel more vulnerable, so that they resisted it both for themselves and for others. Needing a guide altered their relationships with friends – they might choose not to go out with friends because they did not want to impose guiding responsibility on them, and they could not go without (see chapter 12). Driving was a particular feature which distinguished them from others. It made the age of 17 key for some – their friends are all learning to drive, and they cannot (Bethany). This might be for them the first point at which they realise that Usher is going to make a real difference to their lives, as Alex said. They also become dependent on

others - changing the interface of their relationships because they have to ask for lifts, instead of being able to get themselves around.

In general, the participants continued to get around, even if not as much as previously, but what was key in their discussion of mobility was not how they got around, but how it made them feel about themselves.

Giving up a driving licence was not just an inconvenience; it was a disinheritance and the marker of permanent, unignorable change. Driving and having to give up driving, having cane training, or deciding to get a guide dog were significant markers for individuals, and around which they often framed their acceptance and/or frustrations of Usher syndrome. Mobility issues made participants confront their vision loss on a daily basis – which was very unwelcome to some.

7.8.2 Unpredictability

As with other aspects of Usher, the changes in mobility were not predictable. Across the age ranges and the types of Usher, people had lost very different amounts of vision and therefore adopted very different approaches to mobility. Two young people had a guide dog; others had really only just noticed that they could not see very well in the dark. One older person did not want to go out – others travelled confidently. While some aspects perhaps were more predictable – it did not come as a surprise to most participants when they lost their driving licenses (although a shock) - how they would cope, as individuals was not so easy to guess. The unpredictability in relation to mobility was not just in the lack of vision but in their own response to it. They did not know until it happened what they would do. They might think and plan for a guide dog, but they could not predict whether they would reach the point at which they thought they needed one.

There is also little expertise in mobility for people who have hearing loss as well as vision loss. Mobility specialists usually teach strategies which rely on hearing; these are not likely to be accessible to most people with Usher. Farah was lucky to find a mobility instructor who worked well with her instead – this is not everyone's experience.

As with other areas, there is of course unpredictability in the environment as well. The unpredictability which affected them is partly

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the static features such as lighting and things left lying around, but also the unpredictability of public transport (with stranded Rob for example) or the unpredictability of the attitude of taxi drivers (which made taxis difficult for Lucy).

7.8.3 Diversity

Despite their insecurities related to issues of losing mobility, most participants did get around. They used a variety of strategies, depending on the level of their acceptance of aids and support. Some were creative without getting any special support - Megan used an umbrella instead of a cane, Richard had a freedom pass, Susan used a communicator guide to get around. They still travelled, to work, to school and out, although they now avoided dark places, nights and evenings or places where they could not wear their hearing aids.

Their responses to difficulties in mobility were also diverse. Some found different ways of getting around more empowering, particularly those who are guide dog users, or those who found reasons why not owning a car was beneficial in the current age – for financial and environmental reasons. Others felt more restricted and limited, in particular by not driving – the car was there, on the drive, but Julie felt frustrated that she could not drive it.

8 Communication

Communication is a basic need of individuals, not just to make their needs known, to get them satisfied, but also in relation to human relationships, which make us what we are, and the cultural role of interaction, in significant ways such as accessing election speeches and minor but very important ways such as shopping. Individuals need to speak, but also to be heard. Communication allows people to make their mark on the world.

This chapter will explore communication. We shall begin by looking at face to face communication, before moving on to written texts, then technology including telephones and computers. Finally the discussion section will bring together the themes of change, predictability/uncertainty, diversity in order to look at the place of communication in the lives of people with Usher.

8.1 Face to face communication

The importance of communication, and the fear of being unable to communicate is expressed by Chris:

I used to think oh if I lose my sight life is pretty much over with because without communication, cos if you ask what is life, the answer is communication pretty much, everything you do is about communication, if you haven't got vision you can't communicate through sight, and if you've got hearing loss, there's always the dark side in you that thinks what will be the point in living anymore, because how am I going to communicate? Can't hear the TV. Can't see it. Can't hear my friends on the phone. Can't lip read. (Chris, 33, type 3.)

With regards to their preferred method of face to face communication 31 participants used spoken English, 7 used BSL, 2 used Hands on BSL, and 2 used BSL with SSE elements. Nobody had explicitly changed the way that they communicated because of Usher, except Doreen (53, type 1) and Susan (48, type 1) who had moved from BSL to hands on BSL

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over a period of time. Faith (23, type 1) however began learning Paget Gorman whilst at primary school; it was only when she went to secondary school that she had access to BSL. Olivia (54, type 1) and her sister Gill (59, type 1) were banned from using BSL whilst at school but on leaving school it became their preferred method. Farah (17, type 1) and Rebecca (29, type 1) only started to have access to sign language when they began to receive education in BSL at the age of 7/8. Hamid was very aware of how the communication needs of people with Usher vary from person to person:

My Usher's is not going to be the same as another person that has Usher's. I am hearing impaired and I need font 14. Another person they can't read. They need hands on. They need font 16. They need magnifying glasses. Yes they have Ushers so we have to try and compromise. You need to make aware the different levels. Like the deafness. With the d or the big D, how they communicate, how they sign, again when they sign I have to say please I'm Ushers, sign slowly please. (Hamid, 44, type 2.)

Some participants used additional methods to support their communication, for example Rosie, a spoken English user, explained how she used the deafblind manual alphabet at times:

Cos I use the deafblind alphabet sometimes. Just basic spelling out, not as a main form of communication just if I don't hear something or if it's dark, or in the cinema and what did they say?!! A couple of my friends and my mum knows it. It's good for basic communication.
Rosie (21, type 2.)

Jess (15, type 3), also a spoken English user, used hands on BSL when not using her hearing aids. Hamid (44, type 2) felt more comfortable in a Deaf environment than a hearing environment and was seeking to complete his level 3 in BSL, however he was still primarily a spoken English user. Linda (46, type 1) and Olivia (54, type 1) BSL users who had hearing children also adapted their communication at times using their voice, simplified BSL and gestures for example, as Linda explained:

With my husband we use BSL, because my husband's deaf. But both my children are hearing. So we tend to use more gesturing, SSE, and lip reading. Both my children can use BSL, but they're a bit lazy. They don't sign that much. **(I)** (Linda, 46, type 1.)

Participants also spoke of their parents learning 'bits' of BSL when they were young. Erin (30, type 2) learnt sign language when she went to a residential school for deaf children, "I didn't know many signs until I went to boarding school at 12. Everything changed! It was easy to pick up. But I talked more than signed." As Erin was educated in Ireland she is referring to Irish Sign Language, however now in her relationship with Abigail (29, type 1) Erin uses BSL frequently, and as Abigail's needs are changing they are beginning to use hands on BSL.

Participants spoke of issues with communication that affect many deaf people not just those with Usher. Rebecca (29, type 1) commented,

"My parents are hearing but it's hard. We have communication breakdowns. My mum is regularly writing things down for me." **(I)**

Abigail (29, type 1) and Jayne (28, type 1) spoke of having to get their mothers to accompany them to the doctor. Olivia (54, type 1) was frustrated by typetalk. Many hearing aid users found background music in pubs and restaurants distracting. Rob (22, type 2) was unhappy with the cost a £20 taxi to go and collect his 'free' hearing aid batteries. However participants also spoke of difficulties that are more likely to affect those with Usher.

Not being able to follow a conversation, either signed or spoken, was difficult for many participants. Due to the reduction of the visual field some signers relied on visual frame signing, or slower signing, however when communication partners forgot about this participants either missed parts of the conversation or chose to 'zone out'. Rebecca and Linda shared their experiences:

I need that visual frame signing. Or a little bit slower. Sometimes my friends forget, which is a bit frustrating. At deaf club it's a nightmare sometimes because the signing is all over the place. Some people sign to me slow but other people sign really fast. I give up sometimes, because I don't get the information. I miss out on it. **(I)** (Rebecca, 29, type 1.)

...if they're too close I have to ask them to step back and say that you are out of my frame. Obviously if it's hands on I need to be close for that. I do have to remind people to step back, and you need good lighting. Sometimes if people are waffling on at breakneck speed I just switch off, or perhaps walk away. Because I can't follow it. **(I)** (Linda, 46, type 1.)

Also because of Usher's propensity for change, Linda spoke about preparing for the future:

...deafblind manual as well. My family know how to use that. So I'm ready for that if that happens. Recently I have been getting more tired watching sign language. Maybe I am going to have to think about altering communication in the future. **(I)** (Linda, 46, type 1.)

Likewise spoken English users got fed up and embarrassed in having to ask people to repeat themselves, or just 'gave up' as

...a lot of the time people will be at their computers. Or they may have a beard. Or looking away from you. In the commercial world people don't always have the patience or the tolerance to speak nicely or slowly, or speak to you face to face. Stuff like that. They might do it for 5 minutes. After that they get fed up with it. So it's in your own interests not to wind people up so much. (Steven, 36, type 2.)

Even at work I find it tiring. Because my brain is working overload. It's trying to compensate for my hearing and my eyes. Because when I'm trying to talk to them, when I do try and talk to them, where you do it naturally and don't even think about, with me I'm working out every word all the time. And if they move quickly then I've lost them. I've got other things going in my head, and that's probably what loses the conversation in my head with me. I know what's happening but there's nothing that I can do about it. I need to get them in a good spotlight outside, with no noise and stuff! Plenty of light! (Michael, 34, type 2.)

Listening and concentrating is also hard work as Ryan and Jess explained:

Because I'm always tired as well. From my hearing. I always get home, go to bed, for a couple of hours. Eat my dinner and go back to bed. Cos I was too tired. Doing twice as much. Focusing. (Ryan, 17, type 2.)

[Teachers] just tell me to do good listening. Listen hard. Make sure I concentrate. But when I've been in school and I have concentrated I'm very tired. So then I don't really listen very much. That's because I turn off. (Jess, 15, type 3.)

Although these difficulties may be shared by people with hearing impairments who do not have Usher, the combined effect of visual and hearing impairment can leave people with Usher with higher levels of fatigue and exhaustion (Wahlqvist et al, 2013.)

Participants were less likely to report difficulties in producing speech, than listening to what someone was saying, however a few participants mentioned that had had speech therapy when they were younger, or they had words that they found difficult to pronounce.

All adults with type 1 Usher were signers, with only one of the young people with type 1 Usher using BSL. The other four young people with type 1 Usher, had cochlear implants and used spoken English. One person had bilateral implants, another person had bilateral implants but only used one, one person had a single implant but was going to have a

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second implant in a few weeks, and one person had a single implant. As more people with Usher syndrome are choosing to be implanted, fewer people with Usher are full time sign language users. Bethany's and Harry's mother spoke of the distinctions between the 'generations':

And you know this age group forget [BSL]! The majority... The older lot... they were born and there were no cochlear implants available, so they were very in the Deaf world, even like they don't wear any hearing aids or anything, so they're relying on some sort of sign language or hand on hand or whatever; this generation are oral... (Mother of Bethany, 17, type 2.)

The Deaf community don't think people like Jack should be oral, they should all be signing, and live in the Deaf community, and it's **so** not right now. There's some obviously, but a bigger and bigger percentage now can speak, have got some hearing with a hearing aid, don't want to be part of that... (Mother of Harry, 15, type 2.)

These comments also highlight a potential unease between people who have type 1 Usher and see themselves as Deaf and people who have type 2 Usher and see themselves as oral, as identified by Hamid and Victor:

...with Usher we have two groups. One is for sign language users so they are the deaf Usher type 1 Ushers. Then you've got the type 2 Ushers who are orally, now most of them are called the Hearing and Sight Impairment. But with the two groups it's a bit like Manchester United and Manchester City. They hate it each. They can't get together as one person. Oh we sign, oh we can't sign we lip read. Oh give up and walk away! (Hamid, 44, type 2.)

If you have Usher 1, it's a different group, there's some other kind of group, because they are born... they rely on sign language. It's different to us. They communicate very differently. We're in the hearing world and they are in the deaf world. (Victor, 56, type 3.)

For the majority of participants who made distinctions between the communication methods of different people with Usher it was less about how these antagonisms were played out and more about the practical

difficulties of people with different types of Usher and methods of communication getting together. Nicola, Steven, and Doreen explained:

And they come up to you and start signing to you, and I feel a bit ignorant because I don't sign. I've never learnt it. (Nicola, 23, type 2.)

The thing is with Usher people, you get a lot of people that wouldn't be able to communicate with me. Or I wouldn't be able to communicate with them more. Cos they sign and I don't. (Steven, 36, type 2.)

I've been before but they could talk, there weren't a lot of people like me – deaf from birth. Some of them went deaf later in life and have got RP... lots of them do lip reading. A lot people who have Usher are just partly deaf and lip read. If they sign we can use hands on. (I) (Doreen, 53, type 1.)

8.2 Written communication

As with face to face communication participants accessed written communication in a variety of ways. Only three of the participants were braille users - Jess (15, type 3), Victor (56, type 3), and Susan (48, type 1). Jess and Susan were fluent braille users, whereas Victor used braille for labels, preferring to use a (human) reader to access other forms of written communication. Doreen (53, type 1) used a handheld electronic low vision aid, and a small number spoke about handheld magnifiers and task lamps. The majority of participants preferred standard sized print but some preferred larger font sizes, 16, 18, going up to 36. Some participants preferred using coloured paper, usually yellow, and different coloured inks such as blue or purple. Younger participants were more likely to select a preference for different coloured paper and ink.

As well as the three people who used braille regularly, three people had learnt or were in the process of learning braille. Participants found it satisfying and fun, and built up their self confidence as Michael and Rob explained:

And the braille I did at college. It was part of my course. I didn't have to do it but they introduced me to it and I loved it. (Michael, 34, type 2.)

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There's a charity round here that's teaching me braille at the minute. And they have been fantastic! They've helped me, they've supported me... They're (local blind charity) absolutely fantastic. And that's the only support network that I've got. But there's only so much they can do as a charity. (Rob, 2, type 2.)

Other participants were put off by the perceived difficulty of reading braille, yet at the same time were still aware that it may be a possibility in the future as Steven and Alice explained:

I wonder if I should learn braille. But I'm too busy to learn it. When am I going to get time? There's bills to pay. I've got children to bath, wash whatever. Great fun. I enjoy it a lot. But at the end of the day I'm knackered. I'm not going to learn to interpret a few bumps on a piece of paper. (Steven, 36, type 2.)

I know braille is going to be an option in the future. But I'm just not ready for it just yet. I just can't imagine like how, like you see it all the time on medicine packs and that, how do you know? It will definitely something I will look into when it's near time. Maybe I will. (Alice, 41, type 2.)

However the biggest barrier to learning braille was lack of access. Sally (42, type 2) had been advised that "braille was dying out" by her local school for the blind so she was unable to get access to braille classes. Doreen (53, type 1) asked the researcher where she should begin to find braille classes. Hamid (44, type 2) had bought the RNIB's self study guides but was unsure where to start. Carolyn (39, type 2) was frustrated that she would be liable for the costs in learning braille, and felt that information on classes should be more readily available.

Learning (or thinking about learning braille) was also important for participants in putting an element of control back in their lives when faced with the uncertainty of Usher, as Linda and Olivia shared:

I have been thinking about learning braille... like should I start doing that now just for fun? Just so I can start to learn to prepare myself for the future. When the time comes. I mean the time might never come. But it could still be fun to learn. **(I)** (Linda, 46, type 1.)

My sister is getting worse and worse and I say come along, you've only got about a year left, come and learn braille. I would be getting myself ready. I'd like better access for people to have preparation. It's my favourite word for the future – preparation. **(I)** (Olivia, 54, type 1.)

Participants wanted greater awareness of the communication needs of people with Usher, especially with regards to medical professionals and staff in hospitals. It was felt that people in these positions of all people should be able to 'get it right'. Also even when they had been requested participants found it difficult to get letters and documents in large print as Rosie explained:

It seems to be quite a struggle to get large print letters, even from colleges and university, I don't get large print letters. Even though I have specified that I need them. The only place that I can remember getting them from was the bank, but anything else no. (Rosie, 21, type 2.)

Jayne (28, type 1) and Abigail (29, type 1) spoke of letting their mothers deal with their mail, with Jayne commenting that she found it difficult to keep up with bills and important documents. However it was unclear whether this was because they had difficulties in accessing the text, or since English was not their first language (they were BSL users) they had difficulties in comprehension. Similarly Gill (59, type 1) did not like using websites to read information about Usher as she found it difficult to understand all the English, much preferring to have information signed to her. Rob (22, type 2) had had to get his mum to help him fill in his passport application which had been a very frustrating process.

Hamid (44, type 2) was a strong advocate in encouraging people with Usher to tell people what they need in terms of communication, for example be clear with note takers what is needed, the size of writing, the

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colour of pen for example. If an individual does not say, their communication needs cannot be met. Similarly participants did not want others to second guess their communication needs, which was often more unhelpful than helpful, as is shown in chapter 10 on education where some participants resented the use of large print documents. Olivia also commented:

Sometimes it borders on being patronising. They are over the top. I went on a training course back in November. Before I went I sent a request for a large font. I didn't ask for anything else. I arrived and a deaf man said oh this is your seat, over here, over here! The man gave me a laptop and said you can look down and read what they are saying. No, did I ask for that? What I prefer to do is sit within the group and watch it and then look at the enlarged print if I want to. If I keep looking at the laptop I'm going to keep missing the rest of the group. It was an assumption. I mean he was deaf but it was a learning curve for him. **(I)** (Olivia, 54, type 1.)

8.3 Technology and equipment

Technology and specialist equipment was used by the majority of participants, however for many participants who used items such as iPhones, iPads, smart phones, and mobiles it was difficult to distinguish between using them for the sake of using them, and using them as an assistive technology. As Nicola (23, type 2) commented, "Since the iPhone, communication has got a lot easier." Linda (46, type 1) reminisced about the old days when she used to send faxes, and Jayne recalled the hassle of using minicomms:

In the past I used to use a minicom but I don't know where that's gone now. It's old fashioned now. Because there's a third person involved it just takes so much longer. It's much easier now just to use text. It's old fashioned. I don't think that I have the patience for that now. **(I)** (Jayne, 28, type 1.)

The developments in communication technology and their impact on people with Usher was commented on by Harry's mother:

The new generation of Ushers are a lot different. All the technology, it's natural to them. They'll ping off messages on Facebook or texts or what have you. You know with the technology, with Apple things, there's so many adaptations already on there, disability features that they can already access things quite easily. (Mother of Harry, 15, type 2.)

Some participants did however require specialist equipment and often cost was a barrier to this equipment, leaving individuals reliant on others as Abigail and Jess's mum explained:

I went to the vision centre and had a look at the different equipment. Aids that I might use like magnifiers. And things to help me read, help me read menus, when I'm out and about. When I'm going to a restaurant. But they're really expensive – about £350 pounds! So at the moment I've been relying on Erin to tell me what's on the menu. **(I)** (Abigail, 29, type 1.)

We've been looking at technology and it's 4 grand for a Braille Note, and alright there's texting but you can't always text, for like appointments. It's like she got a letter from school ring for your results, ring, she can't ring, she'll have to email you. All of the sudden it hit me and I thought what are going to do then. The deafblind phone business. At the moment if she wants to go to the doctors or whatever I just ring up because I'm her mum, because that's what you do, but it gets to that age, when your mum doesn't do that anymore. (Mother of Jess, 15, type 3.)

Furthermore Jess was keen not to mark herself out as different from her peers by using a braille reader, by using ear hooks she would look as though she was using a screen reader.

Kat's (17, type 1) family had done their own fundraising to purchase a touch screen computer and radio aid.

Often low level technology was useful, for example, keyboards with contrasting keys, powerful torches and task lights, flashing cursors on computer screens, and amplifiers on telephones. A few participants spoke about buying their own equipment as the equipment provided by local sensory services was not appropriate to their needs. Sally (42, type 2) for example was very pleased with the small credit card sized magnifier that her friend had found for her on Amazon, rather than the huge magnifier that had been provided for her that did not fit in her handbag. The baby alarm that Nicola (23, type 2) had requested arrived when her daughter was six months old. Rob also explained his difficulties with the local services:

It's really funny about equipment because social services around here don't like giving equipment. So... they don't like giving things out. So I have had a lot of problems trying to get things that I need. I mean the brailier that I've got that's on loan from a local charity, who's teaching me braille at the minute. (Rob, 22, type 2.)

8.4 Discussion

8.4.1 Change

In relating to change, the most significant comment perhaps is that many of the participants did not change even when others might have expected them to. Those who used spoken English continued to do so, even when their vision deteriorated, although they might have used signs in some circumstances (Jess, Rosie) or have learnt some sign language (Hamid, Erin). (people who spoke and listened generally had continued to do so – even the older people still used the same way to communicate)

This is similar to the way in which participants did not, for instance, want to use a cane, until they could no longer manage at all without one.

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Participants who used BSL as a first language, even if they had had an oral education, did not go back to oral language, but instead moved to hands on BSL. Similarly, although a number of participants mentioned braille, most had not learnt it – unless they simply could not manage without. Consistency was a significant feature of people's responses in this area.

The other significant changes in this area were not to do with the participants so much as with the environment in which they live. Cochlear implants had had a strong effect, amongst our participants, in the number of people who might have previously used BSL but who were not spoken English users. This was a **very** definite difference between the young people and the adults, even given the small numbers overall.

Technology was also changing the picture. Most people had found ways to manage their communications, from emails to telephone calls. Although Jess did not want to seem different in her school, as a blind user, she managed to access her phone. The advent of texting and electronic magnifiers had changed things for some people.

8.4.2 **Predictability/uncertainty**

The nature of the visual deterioration in participants – its unpredictability - meant that they did not then know when they might need to use another method. Some people had notched up an alternative - they had started to learn some signs, or deafblind manual, or were thinking about braille. Others were not thinking about these things until they happened (Alice). Not knowing whether they would even need braille for instance, - the time might never come, said Linda – and she was right. This might mean that people left changes up till the last minute, as Olivia said about her sister. Such late learning might make it more difficult – though braille is best learnt by touch if it is going to be read by touch, signs may be easier to access through touch if once learnt by vision.

Also unpredictable was the availability of help. Braille classes were extremely difficult to access and there was very little information about them (Sally, Hamid, Doreen all had difficulty finding out). Brailers and lessons are expensive and there was no help, certainly not before people actually became blind (Carolyn).

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Because they never knew if they would need it, participants were perhaps reluctant to invest a great deal in learning braille, in either monetary or cognitive and emotional terms.

8.4.3 Diversity

Participants described a range of ways in which they communicated, and a range of communication partners. While spoken English was undoubtedly the most common, and was very stable (people continued to use it, even when their vision deteriorated) so people with BSL as a first language continued to use that, with some moving to hands on. Other sign languages/formats were used such as Paget Gorman and SSE. People used braille as well as print and large printed for text materials. Access to technology meant they could have a far wider experience, and better access, even through using standard equipment which already had good access built in (Harry).

9 Family, Friends and Support Networks

Interactions with family and friends were interwoven into the stories that participants told us about their lives and experiences of Usher.

Consequently this chapter will explore those relationships beginning first with family, before moving on to look at parenthood, and then friends.

Despite all the support that family and friends generally provided to people with Usher, the majority of participants spoke of the invaluable support that they received from external support networks, both formal and informal, consequently this will form the fourth section of this chapter. Finally the discussion section will seek to bring together the themes of change, predictability/uncertainty, and diversity in order to look at the place of family, friends and support networks in the lives of people with Usher.

9.1 Family

When talking about interactions with family the great majority of participants spoke of relationships with close family members, i.e. partners, siblings, and parents. In relation to Usher, participants spoke about the support that they received from their family, and some of the challenges that they faced with family members because of Usher.

Participants spoke about the large amount of support that family members provided – both practical and emotional support. Female participants in particular spoke about the emotional support that they received from their mothers when first diagnosed. Although participants were sometimes frustrated by the extent to which their relatives supported them, there was generally an understanding that people wanted to help, especially with regards to partners:

I rely on my husband quite a lot. And I know he wants to do it and everything and he'd do it without hesitation, but yeah I do rely on him quite a lot. Especially with the kids' activities after school. He has to drive here, there, and everywhere. And whenever we go out anywhere he has to kind of grab me, or lead me, or sometime I say I need toilet, can you take me to toilet. Cos you don't like to say to somebody can you lead me to toilet. He's the one that gets the brunt of all that. He don't mind doing it! (Julie, 38, type 2.)

As Megan (24, type 2) stated in a matter of fact manner, her boyfriend supporting her was "the way of life really." Participants also highlighted the fact they too supported their partners, as Faith (23, type 1) commented,

Me and my partner we're just normal partners most of the time. It's not please help all the time.... And he has to ask me for help as well sometimes. **(I)**

Participants also explained how they still did work around the home but that they might avoid doing certain chores such as washing up, ironing, and painting. Julie (38, type 2) also had had a lot of support from her mother but recently her mother had become ill so was no longer able to support her, which also raises the question of what happens when support networks break down.

Participants were quick to suggest that, in relation to partners, if a person was not willing to support an individual they were not a suitable partner for them. As was seen in chapter six on diagnosis Nicola (23, type 2) and Carolyn (39, type 2) were anxious in telling their partner about their diagnosis, however for Lucy (27, type 2) it was just assumed that her partner would take on board the diagnosis. Gill was also pleased to note that 'nowadays' people were more understanding about things such as Usher:

I think people have got a better attitude nowadays, and they've got a much better attitude out there. In the past you wouldn't know what to say and how to say it. **(I)** (Gill, 59, type 1.)

Nevertheless for those people without a partner, and who were looking for one, they were concerned about how to tell a potential date about the fact they had Usher. One person also felt their difficulties in finding a partner were compounded by the fact that they were bisexual and had Usher, saying “I bet there’s not many of us!”

Due to the progressive nature of Usher, the implications of Usher may not initially be apparent and unexpected difficulties may arise as time goes by, as Steven explained:

My wife, she knew about it when we first started going out. But I think she’s having a hard time of it now. Because before it was a minor issue. Now it’s becoming quite major. Especially when it comes to looking after the children. (Steven, 36, type 2.)

However in order to try and understand more about the ways in which Usher affects daily life Barbara’s (50, type 1) husband and Bethany’s (17, type 2) parents had been on a deafblind awareness course.

As well as support from partners and parents, a couple of participants spoke of support from their children. However as Julie and Linda explained, they found it difficult as they felt that they should be the ones that support their children:

But it’s amazing how kids adapt too... my eldest could see that I were struggling, and she just grabs hold of my arm and leads me automatically. Which is nice. But I don’t always want it, because I feel as though they shouldn’t have to do that. But they do look after me. (Julie, 38, type 2.)

One time we went camping and the lights went out, and I couldn’t see at all. My husband was somewhere else, everyone else was signing, the children were very young at the time, we were trying to fingerspell. I had to rely on the children. I’m supposed to be responsible for them, not them responsible for me. **(I)** (Linda, 46, type 1.)

Despite participants' family members being willing to support them, the stress and burden and doing so constantly can have negative effects on family life as Doreen (53, type 1) and husband, and Steven explain:

Researcher: So usually you go out with husband?

Doreen: [Yes it's frustrating because sometimes we do end up having a row. It's inevitable really. **(I)**]

Husband: It takes 5 mins to explain something to her, very, very simple. She won't tell me if she doesn't understand me because she tries to protect my feelings. And when I say to her, what did I say? She hasn't got a clue. So I have to tell her again. If she had told me the first time!!

You do feel that you're not throwing your own weight about. So you try and do little things, but you're knackered because you're trying to pass as normal, or you're trying to think of everything. And you always forget to do things. We have many, many arguments. (Steven, 36, type 2.)

Usher also caused difficulties for some participants, with their siblings. Bethany (17, type 2) for example recounted how she was seen as the 'favourite' among her siblings as she got the most input from adults, which she found very frustrating. Likewise Alice faced difficulties with her sister, who Alice felt did not understand her situation:

My sister, I don't get any sympathy from her. We had a row one time and she told me to get off my backside and get to work. I wish it was that easy. It's not. (Alice, 41, type 2.)

However for some participants Usher was not that much of a talking point between brothers and sisters who did not have Usher as Kat (17, type 1) commented:

Well (siblings) only know that I'm blind and deaf. So they do know I have Usher. So they just carry on, thinking I'm just normal, like everything's going to be ok. (Kat, 17, type 1.)

Carolyn (39, type 2), Steven (36, type 2), and Nicola (23, type 2) commented that the point at which their siblings (who did not have Usher) became interested in Usher was when they were thinking of

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having their own children, and whether or not they were carriers of Usher.

Even for those participants whose sibling/s had Usher not everyone spoke about Usher. When asked whether she spoke to her elder sister (who also had Usher) about Usher, Dima (15, type 1) laughed and replied, “Not really! She too busy.” Michael too did not speak to his brother about their Usher:

Nah! We don't talk to each other like that. We all get on, but we're all quite independent in our own lives. I think we laugh at each other more than anything, if we bump into anything we go ha ha ha!
(Michael, 34, type 2.)

Likewise Erin (30, type 2) and Abigail (29, type 1) a couple, both with Usher, looked at each other and laughed when asked if Usher was a feature in their relationship, after a moment Abigail responded:

We cope well with each other, even though we've both got Ushers. We walk together outside. If there's a problem coming up Erin always lets me know, and I will do the same for Erin as well. We're always mindful for each other and watch out for each other when we are out and about. We have a great laugh with each other. **(I)** (Abigail, 29, type 1.)

9.1.1 Parenthood

Eleven of the participants were parents – 3 were fathers, 8 were mothers. One participant was about to become a father. Two participants were already fathers when they found out that they had Usher, the other 9 knew about their Usher before becoming parents. In thinking about parenthood there were two issues that were particularly relevant for participants. The first was whether participants would pass on Usher to their child, and secondly, how as a parent with Usher they would cope with being a parent.

Many participants were knowledgeable about the pattern and probability of genetic inheritance of Usher, and the low probability that their child would be born with Usher. For the majority of people this knowledge came from consultations with specialist genetic counsellors. However some of the participants did not have accurate knowledge of the

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inheritance patterns and incidence of Usher – for example Dave (46, type 3) on finding out about his Usher had a vasectomy; he felt thankful for having two ‘healthy’ children and did not want to risk having a child with Usher. Dima (15, type 1) and Hamid (44, type 2) who were from Asian families where consanguineous marriages were common, were however concerned about the way in which Usher is passed on. Hamid had been advised by doctors, and his own family, not to marry within his family:

...my mum brainwashed me the same thing back. You’re never going to marry the family! So I never did that. I’m a high risk. If I marry within my own community, my own culture, it’d be higher risk. In my culture, in my community we marry cousins. There’s more higher risk of Usher... I’ve been told to find a not Asian person to avoid the risk. (Hamid, 44, type 2.)

Hamid went on later to discuss how this was easier said than done – a non-Asian person may reduce the chance of passing on Usher, yet at the same time this person is less likely to be aware and fit in with the culture and norms of a Muslim family. Dima was less direct in linking her questions and concerns about genetic inheritance with cousin marriage, however she had spoken about her larger extended family and their Dima also asked the researcher to provide more information on the genetics of Usher and consequently her and her father were signposted to Sense’s information services. The topic of the inheritance of Usher and consanguineous marriages, was one of the very few times in the study when the ethnic backgrounds of participants had any particular bearing on the comments made by participants.

Having weighed up the ‘risks’ of having a child with Usher, or ignoring it as Julie (38, type 2) suggested that she did, a number of participants were keen to suggest that even if they had a child with Usher “it’s not the end of the world” (Sally, 42, type 2). Ryan (17, type 2) commented, “To be honest I’d be pretty proud if my children had the same as me.” Bethany (17, type 2) and Nicola (23, type 2) suggested, as a person with Usher they were best placed to support a child with Usher. Participants were proud of what they had achieved in their lives despite having Usher and consequently did not see that it would limit their child’s life, as Chris explained:

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Baby on the way. I always been well if I pass it on, I pass it on. I wouldn't change where I am now, I'm not saying that I would want my child to go through what I've been through, but it's not stopped me doing what I want to do professionally, it's not stopped me playing sport at the highest levels. If it did have my disability, I didn't see it as the end of the world, or I don't see it why that child, or it becomes a person, can't be successful in something that they want to be successful in. (Chris, 33, type 3.)

Additionally Carolyn (39, type 2) and Sally (42, type 2) suggested that all parents worry about their children, and if the 'problem' is not Usher, other difficulties will emerge.

Participants spoke of the challenges that they would face/faced as a parent with Usher. Communication and seeing/tripping over children was mentioned as being difficult in the early years, and by participants who had not yet had children, as Faith and Erin explained:

It's like when the children are crying, it's like what are they doing? There's always communication issues, and I don't know what they've banged themselves on because I can't hear. I think it would be incredibly hard work if I did. But I'm really not sure. **(I)** (Faith, 23, type 1.)

I think it would be difficult if I had a baby crawling about all over the place and I would have to keep an eye out on the floor because I would obviously trip over. (Erin, 30, type 2.)

For some participants the greatest difficulty was that they were not able to support their partner in parenting as much as they would like. Olivia (54, type 1) missed the opportunity to drive her daughter to places, and wished that this role did not always fall to her husband. Olivia too spoke about the need to focus on herself when going out, leaving her husband to take charge of her daughter, Richard also raised this point:

If I'm out with my wife, I let her, assume her to look after the kids. And I tend to step back which kind of creates arguments, because it's, you know, you should be looking after them as well. (Richard, 48, type 2.)

Steven in particular was not coping so well with the challenges of parenthood and Usher:

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I feel quite bad... It's really, really difficult. Really really difficult. Cos the thing is because what I had before, and what I've got now is crap really. The quality of life... Obviously I'm rich in terms of children. I've got a fantastic wife. I've got my mum and dad and they're great. And my brother's fine. It's just that outside of that my quality of life is pretty rubbish. Things I know I would be doing if I didn't have this. Driving around. Taking the kids out. All three of them. Give my wife a break.There's no positives to it. I like to think that there is a cure. But I don't think that there will ever be one. Because there's not many people like me about. It's like what's the point in sorting me out. It doesn't make commercial sense. Sorry I know it's depressing... (Steven, 36, type 2.)

The challenges were so great for Jayne that she had decided that she would not like to have children:

I don't want children because I know that it wouldn't be fair on the children. If I'm not there for them, or if I fall over and trip on them and hurt them, or if they are trying to get my attention and I can't see them. It's hard to look after myself; it wouldn't be fair on them. **(I)** (Jayne, 28, type 1.)

Seeing the pain that her parents had gone through when they found out that she and her brother had Usher made Alice (41, type 2) decide that she did not want to have children and "go through the same thing."

One participant (Julie) also spoke about the prejudice she encountered in relation to her children:

When I got pregnant with (son) I remember somebody saying to me do you think that you should have this baby, cos of your eyesight, and I was quite shocked at that. Cos I thought I'm sure blind people have babies, and why shouldn't they. I was quite shocked that were the opinion out there. A blind person can't have a baby! (Julie, 38, type 2.)

However some of the participants provided solutions to these difficulties. For example Emily (27, type 2) suggested that she would ask Social Services for help in picking up her children in the dark, Nicola (23, type 2) gets her daughter to wear shoes all the time in the house so she knows if she is coming up behind her and produced a flyer for the mother and toddler group to let everyone know about Usher and to apologise in advance if she trips or knocks their child, Hasan (15, type 1) spoke about vibrating and flashing alarms, and participants who had been parents for a number of years suggested strong family networks, and 'good' husbands.

9.2 Friends

Friendships were an important part of people's lives, as seen in chapter 12 on leisure activities and in chapter 10 on education, Friends therefore had an impact on their lives and experiences of Usher.

Participants spoke of a fine line between the support provided by friends 'as friends' and support that went above and beyond what a friend would do. When asked to 'strongly agree', 'agree', 'disagree' or 'strongly disagree' with the statement 'I like it when my friends help me because of my Usher', Jess replied:

I need one in the middle! Disagree. Cos I quite like doing things on my own. But if I do need help they will help me. But I prefer to do it on my own because I know that I can. Because I prefer to be able to say to someone, oh this is what I need. Than someone going to me, oh do you need this. That just annoys me. I just want to tell people what I need and what I don't need. So it's quite nice when they help, but I don't really like it. Cos I like doing it on my own! (Jess, 15, type 3.)

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In response to this question Ryan (17, type 2) also had a strong reaction:

Ryan: I don't really like it. I don't know why. It makes me emotional.

Mother: It's not necessarily help, it's more support. If they help him it's like you're not doing it, you can't do it, that's when he gets stressed. But if he's getting stressed because he can't see. They just walk up to the side of him, he grabs hold of them, and then he walks off. Help – yes he gets stressy. Support no he doesn't...

Ryan: Yeah (starts welling up).

Likewise Dave (46, type 3) and Sally (42, type 2) commented:

Dave: some of the things you've got to admit that you need help, but you don't want sympathy.

Sally: Aye you want empathy. The last thing that you want is sympathy. Understanding.

Friends may be well meaning, however as Linda recounted, they might not have the skills and knowledge to support an individual appropriately:

Sometimes I have to remind my friends to be Usher aware. Not to yank me. To do it gently. Rather than pull me, offer me things rather than drag me off. Sometimes that can be a bit of a problem. A lot of my friends know that I have Usher but instead of saying would you like to do this, they forget and grab me by the wrist and just drag me. I have to remind them sometimes. **(I)** (Linda, 46, type 1.)

However for some people, especially those who were not living with family, friends did have to take on a more supportive role as Alice explained:

When it comes to cooking I used to burn myself a lot. So now I rely on (best friend) to do most of the cooking now anyway. Cleaning and stuff like. I might put a top on and not see a spillage. (Alice, 41, type 2.)

The support that friends provided was real. For example, in winter, whilst at university, Rosie (21, type 2) would be 'stuck' in her room from 3pm unless her friends came and met her and took her out. Michael (34, type 2) commented that his flatmate and friend had to help him in terms of

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mobility, however he was able to return the favour by doing all the housework, and similar things were seen in people in partner relationships. .

Older participants with more significant hearing and vision loss were more isolated, tended to have fewer opportunities to meet people and sustain relationships. If we compare Susan (48, type 1) who had 53 hours of communicator guide support a week we see that she was regularly able to meet up with friends for coffee, however for Doreen (53, type 1) who did not have communicator guide support the situation was different, as she and her husband explain:

Doreen: [No I don't meet anyone. I stay at home most of the time. (I)]
Husband: It is very difficult for her to communicate with other Deaf people. In the Deaf club she can't communicate with them.

There is indeed some evidence (Kyle and Barnett, 2009, Sturley, 2012) that some Deaf people find it very hard to adapt to the needs of someone with Usher. Doreen is not alone in finding it difficult to maintain friends with people in Deaf club.

Rosie also spoke about the ways in which she had been denied access to friends and forced into friendships with people she would not have chosen to be with, by teachers and staff who only saw her hearing and visual impairment and not the person with individual characteristics:

...they used to put deaf students together to work with them, and it was quite difficult at times to get to know other students. We were like lumped together – deaf club. Nobody else would think about it but it used to bother me sometimes. Because there are people who have got a disability that I just **do not** get on with! (Rosie, 21, type 2.)

Difference from the mainstream, is however what brought together Ryan (17, type 2) and his friend:

Ryan: I was just getting all worried about social stuff going on, quite a while ago. Then the teacher overheard what was going on. And told my mate what to say – social services is coming over and all this shit. I was getting all worried. And then they was alright with it.

Mother: Everybody needs somebody to trust.

Ryan: It's funny though because he's half Russian and half Polish. But he understands me and I understand him. So it's cool.

The majority of participants spoke about the difficulties in telling friends about Usher, the reason for this is twofold, firstly participants did not want to 'burden' people with this knowledge, and secondly Usher is very difficult to explain. In relation to the first point Bethany (17, type 2) commented, "Friends don't really like talking about it. Upsets them sometimes. They don't really know what to say or whatever." Dave also commented:

One Christmas about three years ago I sat down... we always have a New Year party, the lights are always on, I have to see what's going on, I invited everybody round and sat them down and said I have to be honest with you, this thing's not getting any better. This is what I've got. Very emotional. But you know who your friends are. And all my friends vowed to never ever to let me down, they would always stick by me. They'd always do anything for me. It's funny, there was like grown men sitting and crying. But being honest with myself for once. (Dave, 46, type 3.)

Usher is a rare condition, characterised by change, which often makes it difficult to explain to others, as Alex (29, type 3) commented, "I've never really met someone when I've said oh I've got Ushers, they're oh I know someone. No one knows what it is." Chris (33, type 3) humorously explained the difficulties of telling someone that you have Usher, "Well if I said to you oh I have Usher, and you'd never heard of it before, you'd look at me, and say oh I like Usher too! [Usher – a well known singer/actor of the twentyteens] Now I don't say I have Usher because... ah you want to be an usher?!" Alice (41, type 2) commented, "[my friends] just looked at me, what are you talking about you know. It's not easy to explain."

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As has been discussed elsewhere in this report, once participants began to tell others about the fact they had Usher, they were generally more willing to accept it themselves, and to accept support from those friends and move on with the next stage of their life, as Alice said, “I don’t want to hide anymore and it doesn’t get you anywhere.”

9.3 Support networks

The previous sections have explored the ways in which family and friends interact with the lives of a person with Usher, in relation to Usher. In most instances this concerned support. However individuals also received formal support from organisations for blind, d/Deaf, and deafblind people, Social Services, and from initiatives such as Access to Work or financial benefits for disabled people.

9.3.1 Formal support

As Britain’s leading organisation for deafblindness many participants had experience with Sense. Linda (46, type 1) had been supported by Sense for over 20 years, remarking that Sense’s support for DLA (Disability Living Allowance) tribunals had been particularly effective. Victor was also pleased with the support that he had received from Sense:

I get a tremendous amount of support from Sense. The most recent one being the incapacity benefit and employment support allowance. The Sense legal services were they helpful in registering with them and helping me through the system, to be exempt from Employment Support Allowance and be put in the support group which I was eventually. (Victor, 56, type 3.)

However a few of the parents and some of the young people were dissatisfied by the services offered by Sense for young people with Usher. Leah (19, type 2) and Chloe (16, type 2) mocked the fact that events they were invited to were babyish, for example teddy bears picnics and meeting Father Christmas. They considered that Sense provides great support for congenitally deafblind people however the needs for young people with Usher are not met:

Sense (holidays) are always very aimed to congenitally, or disabled in other ways. Not aimed at these guys, who want a bit of action, want a bit of adventure, they don't want to go to a farm, or gentle things that they seem to be doing. And they're missing out those more... (Mother of Harry, 15, type 2.)

We found a gap, there wasn't an awful lot for teenagers. Sense are really good with children, and oldies, and with deafblind with additional issues, but they don't do a lot for Usher. It is the biggest cause of deafblindness and you've had all these conferences for CHARGE, conferences for this and for that, and you've had none for Usher syndrome. (Mother of Bethany, 17, type 2.)

Such comments highlight the opinion that people with Usher form a distinct group within the group of people with deafblindness.

The participants who did not know about Sense, or only in passing, tended to be those participants whose hearing had not been such a 'problem' in their younger years – either their hearing aids provided adequate support or they had a 'mild' hearing impairment. Such participants preferred to look to organisations for visually impaired people such as local blind societies, and RP Fighting Blindness. As seen in chapter 12 on leisure activities, NDCS (National Deaf Children's Society) was accessed by parents of young deaf people, especially in the early days after a diagnosis of deafness.

Apart from general support in coming to terms with Usher, understanding how it progresses, and what will happen in the future, the biggest area in which people wanted to support was in relation to benefits, which of course is a form of support in itself. Alice commented:

I just wish there was more help on the benefits side of it. Because you know I can't even get higher rate DLA and when I try and explain about my RP, try and explain about my Ushers. And it's the same with the doctors, they say get the doctor to write out a report, the doctor just writes out RP, Ushers, cataracts, guide dog, you know. (Alice, 41, type 2.)

Rosie (21, type 2) did not understand how Direct Payments worked, and it was only with her mother's help that she was able to access them. Participants felt that those in charge of benefits do not accurately understand what it means to be a person with Usher, as Brian's (40, type 2) wife commented, "Benefits people don't understand – difficulties getting benefits, just because you can see a little bit doesn't mean that you don't need help." Hamid (44, type 2) was frustrated that his appeal for DLA had been turned down, "because by the rules themselves you have to be 100% blind, you can't physically walk. Blah, blah, blah and all that sort of thing." There was a sense of unfairness in the way that the benefits system is organised with Julie (38, type 2) commenting,

There's other people who've got back problems, and they're on benefits for years. I don't understand it! I don't understand why it's like that. That's frustrating.

Participants also received formal support in terms of communicator guides, readers, personal assistants, and carers. The importance of communicator guides has been mentioned elsewhere but it is worthwhile reiterating how invaluable participants found them in helping them to access their local community. Two participants had formal carers – Nicola (23, type 2) and Brian (40, type 2). Nicola's husband was her official carer, however she saw this only as a temporary measure as "He wants to go back to work. We want to be a normal family. You know he go out bring the bread home, I stay at home look after (daughter)." In order to look after her daughter Nicola wanted a PA, however the local authority were not in any rush to provide support as she has a carer at home, which in turn meant her husband could not go out to work. Brian's wife was specific in the care role that she provided for her husband:

Wife: I'm have carer's allowance for him, but all it is, Brian does everything really, like he says, I really am I suppose what you could call it his voice. If there's a letter than needs writing I usually do it, if there's like phone calls,

Brian: I write it

Wife: yes, he writes it, then I rewrite it, and like phone calls, he'll tell me what he wants and what he wants to say, and I do it in my words rather than his, because sometimes he gets a little irritated and things

like that, to actually look after himself, it's very rare that I have to do something really for **him**.

Rebecca (29, type 1) had a communicator guide and a reader. She was very happy with the support that she received suggesting, "Yeah I have good support it makes me feel more confident. **(I)**" Victor (56, type 3) had support from communicator guides and a virtual reader who would support him with correspondence via the telephone. Sisters Olivia (54, type 1) and Gill (59, type 1) both spoke of having personal assistants, and Rosie (21, type 2) was looking to get one in the future, rather than communicator guides. They enjoyed the freedom that person centred support offered them, Olivia commented,

Well like today I have my PA, she works with me every Friday. Like shopping for food, going to places together, phone calls. It's much quicker. **(I)** (Olivia, 54, type 1).

9.3.2 Informal support

Informal support, from other people with Usher, or hearing or visual impairments was very important for the great majority of adult participants. Indeed as was shown in the chapter on diagnosis, adult participants wanted to be put in touch with other people with Usher, on learning their diagnosis. One of the most common pieces of advice from participants was to get in touch with other people with Usher. Dave (46, type 3) said, "Speak to somebody else that's got it. Don't speak to doctors because all they do is frighten the living daylights out of you." Nicola (23, type 2) was keen to reinforce the fact that individuals have to look for other people themselves, information on support networks is not always readily available, "when you're diagnosed get out there. Find people. They ain't going to find you otherwise." Harry's (15, type 2) mother also advised parents to "find other families in similar situations."

One of the most common ways for people to get in touch with other people with Usher, and with hearing and visual impairments was through Facebook – this may represent a bias due to the fact that a large proportion of our participants were recruited through Usher and visual impairment networks on Facebook, nevertheless participants who had not been recruited via Facebook still accessed the support networks on Facebook. Participants found that Facebook was useful because it

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provided easy access to other people in a similar situation, many of whom provided helpful information as Nicola and Rosie explained:

I've since found out there's another Usher the other end of town. Friends with him on Facebook. I've not actually met him. But I know he's local. But when you're first diagnosed they, one of the first things they tell you, probably shouldn't do, but how rare it really is. And they break it down into numbers. And how many in how many have got it. You're like crikey I'm alone. You know. No-one. Who can I talk to? (Nicola, 23, type 2.)

I've only found the Facebook groups recently... I found the RP society, and got all the other groups on Facebook like the RNIB. And Sense. I would never have thought to have done that before. I'm not a huge fan of Facebook. I've only just started finding out how useful it could be. It is quite helpful. (Rosie, 21, type 2.)

The parents of some of the young people also mentioned the value of such groups – in fact one of the groups through which we made contact with participants was run by parents of children with Usher. Harry's (15, type 2) mother commented, "I'm a member of lots of groups on Facebook for Ushers. They are useful. If you ever need any advice on something, somebody will have the answer."

One of the attractions of Facebook is that it allows people to dip in and out, accessing what they need. Jayne commented:

There's Usher life on Facebook. I don't get involved but I do read the stuff. It's rare for me to make comments, but I do read the information there. **(I)** (Jayne, 28, type 1.)

Participants found an online presence important as it can take a lot of courage to meet other people with Usher as Faith and Gill explained:

And I was teased and teased and teased about being blind. And it took a long long time to be involved in an Usher's group. I know I'm not alone in that respect now, but before I didn't know, I didn't understand. **(I)** (Faith, 23, type 1.)

when I first started and I didn't have the confidence and I didn't know what to do, but as time has gone on it has been lovely to meet other people and share experiences. **(I)** (Gill, 59, type 1.)

However for other participants it was important to be able to meet up with other people with Usher face-to-face, as Harry's mother (15, type 2) explained, "Because you can read loads of literature and it doesn't mean anything but talking to somebody who's got the same, really makes a difference." For Daniel (15, type 2) it was good to meet up with other young people with Usher as 'they got it' unlike when he met up with his neighbours.

In the first instance participants found Usher support networks beneficial in understanding that they were not alone in having Usher, as Faith and Julie explained:

I emailed Usher support and I found out that there were so many! It was quite a relief to find out that I wasn't on my own because it's quite a relief to find out I wasn't the only one. **(I)** (Faith, 23, type 1.)

I think it has helped for me finding other people a similar age and similar stage. And find out how they are coping with it. At first there was nobody for years. I contacted people on the websites for Fighting Blindness and nothing ever came of it. And you felt like you were hitting a brick wall all the time. And I did feel like I were on my own for a LONG time. And I think when first I found (local blind charity), that's when I first felt that I wasn't on my own. (Julie, 38, type 2.)

As participants began to get in contact with other people with Usher (online or in real life) their desire to meet other people more similar to them grew. They might look for others who shared characteristics such as gender, age, ethnicity, and extent of visual impairment. Olivia (54, type 1) and Doreen (53, type 1) were interested in meeting other women with Usher. Vicky (44, type 2) and Steven (36, type 2) were interested in meeting people who were working in a commercial environment. Steven wished that there was a support network available for his wife. Richard was keen to meet someone around the same age, and with a diagnosis in later life:

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I must admit, three years ago when I was diagnosed with this, I was trying to find people who were similar to me. I went to a forum up the centre a couple of times. Met a whole load of people and there wasn't anyone like me. There wasn't anybody in their forties who had been diagnosed when they 45 say. (Richard, 48, type 2.)

Jess too wanted to meet people her own age with Usher

... I'd like to meet more people my age. And a bit similar to me. I don't really know many, like younger ones. It's nice to meet people but I don't really know anybody my age. It's quite nice to meet the adults because, like (adult) who can tell me everything, it's quite nice because he remembers what it was like when he was younger. So that's quite nice really, but there's no one my age. (Jess, 15, type 3.)

Hamid (44, type 2) spoke about his struggle to find other Asian people with Usher, "trying to meet people, I was the only Asian. I'm thinking where are they. Hello?! Well it can't be that difficult."

Some participants did not want to meet people who were old or whose vision loss was greater than theirs, as they did not want to see what the future may hold in terms of vision loss, as Lucy explained:

I am aware that I don't want to meet loads of people that are older... I know it sounds really weird, but I don't necessarily want to see how bad it can get... And I don't like to think of myself as getting blind, even though I know that it's going to happen, and all that. So to be honest I don't really think about the future that much. So I think that was one of the problems because you start to see, you could end up being that blind. But people my own age that's fine because we can talk about things that [are] current, problems that you face now. (Lucy, 27, type 2.)

Ryan (17, type 2) also said that he did not want to meet other people with Usher "If they have it bad, if they have it worse..." However some participants found it useful to get in touch older people and see how their Usher had progressed and compare themselves. Barbara (50, type 1) spoke of being in touch with a person 10 years older and using her friend's experience as a guide for the future. Nicola explained:

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How am I going to know what's gonna happen in 10 years or whatever. So thanks to the world of Facebook, I've got Ushers in their 50s, 30s, 40s, teens, 20s, so I'm literally seeing every stage, I can imagine what I'm going to be like in 30. It doesn't scare me anymore. Cos I'm seeing they're coping. (Nicola, 23, type 2.)

Participants wanted to gain positive experiences from Usher networks and consequently did not want to focus on the negatives of Usher. As Vicky (44, type 2) commented, "you don't want to sit there all doom and gloom, you just want to get on with your life." Steven shared the same opinion:

(RP Young Professionals) I think that's more up my street actually. I don't really want to go to a sob story Usher gathering, with a bunch of sad people talking about their problems. (Steven, 36, type 2.)

Finally despite the obvious support and benefits of Usher groups, one or two participants disliked the 'everybody knows everybody' situation due to the fact of a small population size, and this could cause difficulties if there were disagreements as people's access to supportive networks could be denied.

9.4 Discussion

9.4.1 Change

Family are one part of people's lives which to some extent cannot change – a parent remains a parent and a sibling a sibling. But once Usher had been diagnosed, these stabilities were moulded by a different set of criteria. Brothers and sisters sometimes watched each other, to see how they were 'coping' (Alex, Lucy). They watched a sibling, or a partner go through change and they wondered what they would do, or they decided to do things differently.

Both family and friends changed their role as participants' visual impairment progressed. This change was a difficult one for the participants to manage, as Jess's conversation suggests. They knew they needed help but they did not want it to have to be their friends. On the other hand, they would not have wanted a 'helper' in those situations either – no-one wants to go to a party with a communicator guide. Their

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family and friends also needed to change, to understand what was needed for someone with Usher – and some made this very formal, for example going to classes (Bethany and Barbara). For others, it was a sea change in the way that friends related to them (as Dave's experience suggests). Wives and husbands, and fathers and mothers, as people with Usher had to accept that the role model they had previously aspired to for their relationships might have to change – they might not be able to drive their children to events (Olivia), or they might have to depend on children to do things for them (Julie).

In terms of wider support, some Deaf people with Usher felt that Deaf clubs were not very supportive, which does link to other research and opinion in relation to this. It may be that with a change in culture in Deaf clubs to be more inclusive, people with Usher would feel that they had more access and more friends; that they did not need so much individual support.

As visual impairment progressed, participants sought a variety of more formal support. This might initially be by browsing through internet sites, but they might then turn in more formal ways, as their need to adapt grows, to specialist organisations, or to specialist networks.

9.4.2 Predictability

One of the key issues in relation to predictability and uncertainty was related to genetics and having children. While most participants seemed clear about the probabilities (although not everyone was clear), and in one sense it was entirely predictable, in terms of statistics, how likely they were to have a child with Usher, in other senses it was a lottery. This uncertainty made Dave decide to have no more children, and Hamid to seek a marriage partner outside his family – a very difficult decision for him. Other parents, knowing that there was a risk, decided to look on the positive side – it was uncertain, and anyway, they might be a good parent for someone with Usher (Sally, Chris, Ryan and others). Less predictable was how they would cope, and how this might change as their visual difficulties increased - there were worries about falling over small children (Erin) and about becoming dependent on them (as above). As with all parents as Sally said, there will be issues. They cannot be predicted for any child. There will be some for a child with

Usher, or a child whose parent has Usher, but they might not be related to Usher.

There was a feeling that support was not very predictable. Some people had excellent support - Susan had 53 hours of communicator guide support a week, which enabled her to get out – but others did not get much support. They found that interactions with, for example, benefits departments had unpredictable outcomes – although they might have had a very good case of support, they might have to fight to get it (for example Victor and also Susan with her bus pass in chapter 7). They might have been quite clear about the type of support they wanted, but this did not mean they would necessarily get it. Nicola did not want her husband to be a carer – they both wanted him to go back to work.

9.4.3 Diversity

Participants showed, as in all other areas, a variety of responses to families, friends and support. Some were happy to embrace Usher within this – to have a partner with Usher, to talk about Usher with siblings – whereas others were more likely to be too busy to do so (Dima's sister). Some were happy to accept informal family support as part of family relationships –as Julie and Faith were – others worried much more about this – Nicola and Carolyn suggested their husbands might want to leave them (in chapter 6). While some did talk about Usher in families or relationships, others rowed, or simply did not talk to a brother or sister at all. One reason for this might be the unpredictability of Usher –what a brother or sister – or partner – had might not be the same, anyway.

Some had good friendship groups and were connected with others with Usher, with Deafness, as well as without (Rosie, Ryan, Alice), while some, like Doreen were much more isolated and alone.

Throughout, participants found a range of support and companionship from others. Some found this in family members, who were understanding about Usher, some found it in friends, who took Usher on board. Some found it in support groups, online or face to face. Others found support in professional help – support that they paid for, but support nevertheless. Their need for both people to talk to and for help

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was therefore gained a variety of ways. Those who were more alone, they shared difficulties in getting out, and difficulties in communication.

10 Education

This chapter will explore the topic of formal education. The changing nature of Usher syndrome means that many of the adult participants did not know that they had Usher whilst at school, so the responses here will be weighted more heavily to include those from the young people (14 – 20 year olds). For the most part participants spoke about their secondary school education; however we shall also include the experiences of those who attended college and university. Whilst recognising that education does not always take place in a school context, the responses from participants centred on experiences at school, so this will be the focus of this chapter.

This chapter begins by exploring the types of schools that participants attended. As well as observing the variety of schools, we shall explore the ways in which the type of schools that participants attended helped shape (or not) their identification as d/Deaf, Usher, or visually impaired. We shall look at some of the specific educational challenges for students with Usher including, access to the curriculum, career choices, and specialist support, as well as sharing some of the successes. School is not just about education, especially for students at residential schools, therefore we shall examine participants' comments on their interactions with peers. Finally in the discussion section, through the themes of change, predictability/uncertainty, and diversity we shall look at what is special about the Usher experience of education.

10.1 Types of school

The type of school that a child attends has a strong impact of the kind of education that a child received and this is particularly so for students with sensory impairments. Möller and Danermark (2007) , writing about deafblindness, not only children with Usher, note that for deafblind pupils there were a number of school changes and that pupils were not always able to be in the schools they wanted or the sessions they wanted. School type can have an impact on the method(s) of communication that a young person uses, the way that an individual sees themselves, and how others see them. For children at residential schools in particular it

impacts upon free time and family arrangements. The participants in the research project attended a range of different types of schools – local mainstream schools, residential schools for the d/Deaf (as boarders and day pupils), local fee-paying schools, grammar schools, local schools with resource centres for hearing impaired children, schools for children with learning difficulties, and residential schools for blind and visually impaired pupils (as boarders). This rich variety of schools that participants attend, or attended as children, highlights the very great differences in the individual experiences of people with Usher.

As was discussed in chapter eight on communication, we see how the communication needs of the Usher population have changed over time, particularly given the introduction and availability of cochlear implants. This in turn tends to have an impact on the preferred methods of communication of people with Usher, which then impacts upon their education choices and the types of schools that they attend. For instance, all 12 adults with type 1 Usher were signers – BSL, SSE, and hands on signing – and all attended residential schools for deaf children (7 as boarders, 4 as day pupils, and 1 flexi-boarding). Of these, the 6 older adults (36-59), although they were signers in adulthood, attended schools which adopted an oralist approach to education. Of the 6 younger adults (21-35) with type 1 Usher, 3 attended schools where sign language was the main method of communication, whereas 3 were at schools that had an oralist approach. However of the 5 young people with type 1 Usher only one was a full-time BSL user and attended as a day pupil at a day and residential school for deaf students and hearing students with language, communication and/or auditory processing difficulties. The other 4, as a result of cochlear implants, were English language users. Two of the 4 attended mainstream schools, one attended as a day pupil at a residential bilingual school for the deaf, and one, who also had learning difficulties, attended a school for children with sensory impairments and additional needs, boarding one night a week.

Of the 16 adults in the research project with Usher type 2, only four were diagnosed with Usher whilst in full time education. Of the four people who were diagnosed with Usher whilst in full time education one moved to a residential school for Deaf students and began to learn Irish

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sign language to support her learning, two attended mainstream schools, and one attended a school with a resource centre for children with hearing impairments. Of the other twelve adults, nine attended a mainstream school, one attended a local school with a resource centre for hearing impaired children, one attended a special school for children with learning difficulties (although he felt that he did not have learning difficulties and it was because of his hearing impairment and communication difficulties that he attended that school), and one attended a school for Deaf children for his first few years of primary education, then moved to a mainstream school, then moved to a special school for children with learning difficulties. Again this person felt that he did not have learning difficulties but because of his (undiagnosed) visual impairment he was not able to keep up with classmates in a mainstream environment. Rob explains his situation thus:

I went to mainstream school but I had problems at the school. Unbeknownst to staff and other people I was blind at the time and I couldn't do my work. So because nobody detected it I ended up getting sent to a special school, where they tried their best, well... I did as well as possible but I felt that I could have done more. I was also at the age where I was "sod school". School was a bit touch and go. I had no support at school. No teachers suspected anything, even though I kept walking into tables, chairs, bags. And I was sat at the front of the class and I couldn't see the board and nobody even picked it up. 18 years nobody suspected a thing! But I just thought it was normal. (Rob, 22, type 2.)

Of the six young people with Usher type 2, four were attending a mainstream school or college, one was at a mainstream school with a resource centre for children with hearing impairments, and one had begun secondary school in a mainstream school, then moved to a residential school for deaf children where teachers used an oralist approach but the majority of students used BSL outside classes.

The two older adults with type 3 Usher did not know about their Usher whilst they were at school and attended mainstream schools. The young adults with type 3 Usher did know about their Usher whilst at school – one attended a mainstream school, and one attended a variety of different schools. Although a long extract, it is very powerful to hear

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Chris's educational journey through the schools that he attended. The issues that Chris raises in relation to bullying and 'fitting in' are discussed further in section 10.4 interactions with peers.

Where do you want me to start?! Ok... I went to a primary school at the end of my road, which is no longer there, but that was a mainstream infant and primary school. But then my hearing started going waywards, so I went to another school which was a primary school and infant but it had a deaf unit. Then my sight went. So then, as far as I'm aware the council said you're better off going to a visual impairment-blind school. Then I went to [residential VI school]. I was there for 3 years... The secondary school for the blind and visually impaired said you'd be better off in a deaf school. So then I went to an oral [residential] deaf school. The problem was I got there in year 10, and started my GCSEs and imagine how much of an outcast you look when everyone's been together since the start of secondary school, some new guy comes in and he's got a visual impairment – whoopy do!! But I got to admit I probably got a better education through going there but at both schools there was a lot of bullying. One you were bullied because you couldn't see, and then you were bullied cos you couldn't hear – so you couldn't win basically! Then after that I went to sixth form at [residential oral school for deaf children]. Then I went back to visually impaired college, then eventually I went back into mainstream college, then mainstream uni. So I've done a lot! (Chris, 33, type 3.)

Chris's experiences with his changing vision and hearing during his schooldays highlight the fact, as seen in chapter 1 on diagnosis, that the standard textbook understandings of Usher need to be reviewed. Traditional understanding suggests that the hearing and vision loss of Usher type 3 is not experienced until individuals are in their 30s. However both Chris and his brother Alex's type 3 Usher was identified by genetic testing in the National Collaborative Usher Study which began in their primary years. Furthermore we can see that the way in which Usher syndrome is expressed varies greatly, even between two siblings who share the same genes: Alex was able to continue his education within a mainstream environment, whereas Chris was moving

through different types of schooling as his hearing and vision deteriorated.

The young person with suspected type 3 Usher also attended a variety of different schools beginning at a primary school with a resource centre for hearing impaired children, then moving to an oral residential school for deaf children in year seven, before moving to a residential school for blind and visually impaired children in year eight.

Attendance at a particular type of school is generally based upon parental choice. However, funding issues can prevent, or slow down a child attending a particular school. Emily (27, type 1) spoke of her mother going to court in order to provide her with the opportunity to board at her residential school:

I really enjoyed boarding. Well I only just lived down the road from my school, so my council couldn't let me actually stay in the boarding part. So my mum had to go to court. I think... [it]1-2 years to fight for me to be a boarder, obviously because of the expense. In the end my mum won, because she really wanted me to be included in the school community. Not me having to come home all the time. She wanted me to be included. I'm **so** glad she fought for me, because I **loved** boarding. And not having to go home all the time. **(I)** (Emily, 27, type 1.)

Kat's mother spoke about the tribunal process that she was involved in, in order to get the most appropriate education for Kat. As we saw in section 1.3.1 on specialist support, attendance at a mainstream school does not mean that a child is without specialist support. However, for the majority of students this support was from a visiting teacher of the deaf (rather than an MSI teacher) or even a teacher of the visually impaired. Furthermore not all schools in the same 'type of school' category are equal. For example Vicky (44, type 2) and Richard (48, type 2) spoke about their parents specifically choosing smaller secondary schools for them to attend.

Even though a particular school may be the most appropriate in terms of education, it also needs to fit into a child's home life. Rosie (21, type 2) and Hamid (44, type 2) spoke about the sense of isolation in having to attend a school with a resource centre for hearing impaired children and

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the long taxi journeys that were a part of this. Kat (17, type 1) and Bethany (17, type 2), as well as Emily in the quotation above, spoke about the importance of fitting in at residential schools particularly for day pupils, and especially when friends and classmates are involved in social activities in the evenings at school. Lastly, the type of school that a child attends may involve difficult times for parents as well. Jess's (15, type 3) mother, reflecting on Jess's time at boarding school commented: "So I thought that I'd get used to it, but 5, nearly 6 years in I've still not. The only difference is now when I leave her at school I don't feel as though I want to cry all the way home."

10.2 Shaping Identities

The types of schools that children attend play some part in shaping young people's identities. As will be shown in subsequent chapters on leisure activities and work, and the previous chapter on family, friends and support networks, school is not the only arena in which identities are formed. Nevertheless, for the participants in this project the types of schools that participants attended shaped their identity, particularly in relation to their sensory loss - d/Deaf, hearing impaired, blind, visually impaired, or 'normal'. Rosie explains how being at a school for children with hearing impairments encouraged her to see herself as deaf:

Probably one of the reasons why I identified more with being deaf, for years, because I got told that I was deaf. I introduced myself as deaf at primary school, then at secondary school I went to a school specifically for hearing loss, well mainstream school with a specialism for hearing loss, it's probably one of the reasons why I identified more as being deaf, and my vision was just an added thing on the side. (Rosie, 21, type 2.)

Rosie goes on to say that it was not until she started needing a bigger font to read whilst was at university that she began to view herself as blind. Michael speaks of the way in which it was not until he attended a college for visually impaired people that he began to identify with people with visual impairments:

I didn't want to look different really. I just tried to be... I normally get told off for saying this word, but I'm going to say it anyway, I try to be

as normal as possible. (Deafblind worker) always tells me off. I just try and pretend there's nothing wrong with me. But now since I went to college, which I started in January I accept it more. Not the hearing but the eyes actually. The hearing I've got used to anyway... It's more harder hanging out with people who have no visual impairment, because they don't really understand. They tend to leave you behind. They don't mean to, it's their natural way of doing things. Because I came to (college for blind people) and I met some friends and they are VI. It's almost like the army – wouldn't leave a man. (Michael, 34, type 2.)

For Harry (15, type 2) his vision and hearing impairments were not a strong feature of his identity “I don't tell them I'm blind” he said. “We just get on with things” explained his mother. His mother attributed this to the fact that Harry attends a high achieving grammar school, where the significance of his hearing and visual impairments are lost on the teaching staff:

I think it's just because they think he's normal. It's a grammar school. A very high achieving school, they expect everybody to be just work, work, focus, they don't really get the special needs. All special needs. Because there's such a low percentage of special needs children. (Harry's mother, 15, type 2.)

Furthermore identity is not static. Olivia attended an oral residential school for deaf children. Because of the way that she was treated by fellow pupils and also, as she explained earlier in the interview, she was forbidden from using sign language, it was only after she left school that she began to see herself as Deaf and having Usher:

Secondary school, I did keep it quiet [that I had Usher]. It was difficult. I didn't like the label being deaf either. I didn't say anything, and I didn't like signing. I didn't sign in public, I was oral. I didn't like the label. But now I sign. I changed attitudes now. I now tell people [that I have Usher]. Back then I didn't. At the school the pupils knew I couldn't see and they would mock me. **(I)** (Olivia, 56, type 1.)

Difficulties arise when an individual does not feel as though they fit in with the identity put upon them by others, or there are barriers preventing them identifying with a specific group. As mentioned previously, methods of communication can be one way in which an individual associates or disassociates themselves from a particular group. For example, Nicola (23, type 2), who had attended a mainstream school but then attended a college for Deaf students, spoke of being *in* the Deaf community but not being a part of it because she could not sign. Bethany, who is a spoken English user, spoke with an intensity of feeling on the difficulties of fitting in at a school where the majority of other children use BSL:

So starting at [residential school for deaf children] was a little bit alien to me because I was used to the whole mainstream thing. They kind of looked down their noses at me because I used my voice, and I was very oral, and I didn't sign. But I picked up sign language to communicate with everyone, part of me trying to fit in. They weren't you know, sort of, very like welcoming with open arms. It was very much. No, no, you've got to learn to sign. You can't use your voice. (Bethany, 17, type 2.)

Jess's experiences of being a pupil at a residential school for blind and visually impaired children demonstrates the fact that identity is multifaceted and that individuals can adhere to multiple and different identities simultaneously:

When we're like inside the school fence, we're normal, but when we step outside we're a bit weird. We are! Honestly, when we're inside school, playing like football or on the bikes to each other we're normal, but then we go outside people stare at us. If we go in the minibus with school for the blind written on the side, everyone just stares. (Jess, 15, type 3)

Lastly, participants did not speak about the way in which the type of schools that they attended helped to shape their identity as a person with Usher. As we have seen in chapter nine which explores friends, family, and social networks, it is only when individuals began to spend time (in real life and virtually) with other people with Usher that they begin to associate (or disassociate) themselves with an identity of Usher. Instead, for the small number of participants with significant hearing and vision loss whilst still at school, as a result of the fact that they moved between different types of schools, or that the school that they attended did not adequately fit their needs, there was a sense of frustration and a feeling of difference. Jess's (15, type 3) mother remarked, "It's a funny predicament for you isn't it Jess? A funny place to be. Because you're neither here nor there." Chris also commented:

I think the biggest challenge was... mainstream was ooh how do we deal with this child, when I went to a specialist school found that they dealt with one side of the disability but then they weren't quite sure how to deal with the other. (Chris, 33, type 3.)

10.3 Educational Challenges

Participants spoke about the particular educational challenges of being a person with Usher/deaf at school. Although some participants spoke positively of their experiences, a lot of participants (and their mothers) were annoyed, frustrated and upset with aspects of their/their child's education. In particular, dissatisfaction surrounded specialist support, access to the curriculum, and careers advice.

10.3.1 Specialist support

All participants received some degree of specialist support, whether it be a visiting teacher of the deaf – twice a year or once a week, a teaching assistant in all lessons, or support for particular lessons. Students at schools specifically for deaf or visually impaired pupils were taught by teachers specialising in these fields. Nobody spoke specifically of having support in school from a specially trained teacher of deafblindness; however one of the teachers at Jess’s residential school for blind and visually impaired students was training to be a teacher of the deaf.

There were some encouraging experiences of participants’ experiences at school, for example Erin (30, type 2) was pleased that she moved to a residential school for deaf children as “the teachers would know not to move about.” Megan (24, type 2) was also positive about her experiences, explaining how her peripatetic teacher of the deaf would focus on informing the subject teachers the best way of supporting her, rather than “isolating me too much.” Ryan (17, type 2) too was satisfied with the teachers and staff at his school, “All the teachers that I’ve got, they understand what I’ve got. The main teachers that I have all understand. All the important ones know about me and understand. And all my TAs.” Likewise Abigail was enthusiastic about the support that she received at school:

I always used to sit by the window, the school actually helped me and benefited me because everybody was kind, and that I could see the board properly, the black board and the white board, if it was on a blue background I couldn’t see it so I used to ask them to change the colours. They always used to adapt it to my needs. And they used to use A3 paper so I could do my exams using A3 paper. They did help me. **(I)** (Abigail, 29, type 1).

Participants were keen to note that it was often small changes that were of great benefit whilst at school. For instance Farah (17, type 1) found it very useful when her PE teacher suggested using a yellow shuttlecock in badminton, which meant she could continue playing her favourite sport. Julie (38, type 2) and Bethany (17, type 2) spoke of the benefits of being able to be seated in the optimum position in the classroom. Many of the younger participants reported that different coloured pens (blue,

purple, not red, not green) and paper (yellow, buff, blue) helped them to access written materials more easily. Harry's (17, type 2) mother mentioned that it was useful for Harry to be able to wear a cap on bright sunny days. For Bethany (17, type 2) and her mother, a positive can-do attitude was very important:

Mother: ...the college Bethany is going to go to, besides the fact that they are very, very willing, and I think attitude is what it's all about, if you're willing to give somebody a go, Bethany deserves the same as everybody else – "we're happy to learn, you've got to realise that we might make mistakes" - well I prefer that, than we know it all and then making her feel so awful, she can't even go any more. But you know, even at your interview they were so positive weren't they, when they spelt everything out about the courses, and armed with those A-levels Bethany, you can go to... and reeled off all these universities and I thought, I've been through this with my boys, I've never had this before, because they stayed at sixth form, but yeah it's all about attitude.

Bethany: I'm looking forward to it!

Participants also spoke of their positive experiences at university with regards to support, although Rosie (21, type 2) and Lucy (27, type 2) suggested that people's intentions were often better than the outcomes. Participants also spoke of the importance of recognising their own needs and advocating for themselves with Rosie making the comparison between school where support was provided and university where she has to ask for it, and the challenges she had faced in her learning how to do this without the support of her mother. Ben also reinforced the point of asking for the help that you need:

If support and guidance is what they need that's up to them. If they want that independence they can always say look I don't want , or I want this. You have to ask for what support you want. At university you do have to let them know. You're obviously free to go and do whatever you want at Uni, but you have to let them know what you want. You can go out if you want to. You can have a guide that goes with you if you want to. I don't know about social events... you just need to stick up for yourself really and say what you want. **(I)** (Ben, 30, type 1.)

Despite these positive experiences, participants spoke of the problems associated with specialist support. Participants spoke of a desire of not wanting to be singled out and made to feel different from their peers as Julie (38 type 2), and Chloe (16, type 2) and her mother explain:

Even though I didn't have any hearing aids or anything, I still felt singled out when they took me out of lessons, and asked me if I were ok. The visits were probably only once every six months, but I felt as though they were coming all the time. I think just make things easier for yourself, sit at the front if you can. You have to find what's easier and try and find and do what's acceptable for you. (Julie.)

Mother: Well all [the advice] we got told, and she wouldn't adhere to it anyway, is, she has to sit in front of the board. So she can see it. And she's close so she's not missing any information cos of her hearing. She won't do that.

Researcher: Why?

Chloe: I just don't like sitting at the front of the class.

Participants also spoke of the fact that they did not want the extra support provided to them, or that it was not the most appropriate for their needs as Faith and Nicola explain:

For years and years I had to have these A3 pieces of paper and basically everyone was laughing at me. They gave me a tiny wee magnifier. They were trying to help – but A3, I didn't need A3! I could read A4, it's just that I couldn't read what was on the board. About a year later I blew up about it and told them, this is wrong, this is wrong, and things started to change. They were asking me, what would you like? That was much better. I said, can you not do it on A4 please. **(I)** (Faith, 22, type 1.)

So while everyone was in the bulk of the class, sitting together learning from one teacher I had a teaching assistant next to me, nine times out of ten. Relittrating or helping me. "I can do this!" Leave me alone! I'm deaf but I'm not simple. I just struggle. I know they are there to help me but in my opinion, you're not gonna learn until you have to. And I never really had to. It was only really say when I was 14 that I said I **don't** want a teaching assistant this year. And that's when I hit my [age appropriate reading level]! So it was because that was took away from me, I said no you're singling me out, I wanna make friends, you're making it hard for me, I don't want this special attention, and that's when I learnt. (Nicola, 23, type 2.)

Likewise Rosie (21, type 2) and Chris (33, type 3) spoke of the fact that although teachers were informed of best practice, unless they actually act on these recommendations, the difficulties still remain. For Chris these difficulties were exacerbated due to the fact that as a student he would meet a number of new teachers over the years:

Some teachers were just rubbish full stop. You can say to a teacher make sure that you are looking at me when you talk do this, do that, but they still face the blackboard, or whiteboard and talk! They forget... I had a note taker and a communication support worker, they were always were trying to find something to help assist me but really to be honest it's...new for them, and you don't have the same teacher for all your school years. You move up through the different terms and you get different teachers. The things is one teacher might be really understanding and you think great, but then 8 months later, see you later you're on to another teacher. Then you're starting from scratch again. (Chris, 33, type 3.)

Chloe (16, type 2) and Leah's (19, type 2) mother was fed up with having to ring up the school to remind teachers to provide written information in the appropriate format and being met with the response that the teacher must have forgotten. She was also concerned that the number of hours of support that her younger daughter was receiving was reducing, especially when compared with the experiences of her older daughter only a couple of years previously. Jess (15, type 3) and her mother also remarked on the limited number of specialist hours support received:

Mother: Then what happens every year at your annual review? Who comes to that?

Jess: [woman] from Sense.

Mother: And she's a teacher of the multi sensory impaired. And every year what does she do?

Jess: Says the same thing.

Mother: Yep! Which is?

Jess: I need more help.

Mother: Yep and what happens?

Jess: Nothing!

Mother: Yep!

The biggest challenge faced by young people with Usher was making sure that their needs, in relation to both their vision and hearing loss were met. Chloe (16, type 2) and Leah's (19, type 2) mother

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commented, “They’ve never had support for their eyes.” Harry (15, type 2) and his mother were frustrated that those providing support for Harry did not fully understand his needs in relation to his vision:

Harry: They just get one of the TAs to go to the photocopier and enlarge it, but that doesn’t really help because it goes all thin and quite blurry.

Mother: It’s the whole understanding of how the eye sight works. Even though we try telling them all the time. It’s all in one ear out the other.

For example Jess had had many difficulties at the oral residential school for deaf children that she attended in year seven, particularly in terms of her safety and mobility, and being bullied because of her visual impairment, yet when she moved to the school for the blind it took 3 years for them to provide any support in terms of her hearing loss. Particularly poignant however was her mother’s incredulity at the fact that support for people with Usher is so difficult to find within a deaf environment:

So that’s when we went down to [oral residential school for deaf children] but considering only deaf people get Usher, [school] are just rubbish! They really, really are rubbish at it! As in the VI side of things. I just get my head around that a deaf school can’t do the Usher thing. Only deaf people get Usher. It’s not as if you’re blind first then deaf. You are **deaf**, and then the Usher, the VI comes. (Mother of Jess, 15, type 3)

Similarly Bethany’s (17, type 2) mother said, “Nobody understands [Usher syndrome] and nobody seems to want to. We found that with schools for the deaf, they can’t get their head around the deafblind.”

10.3.2 **Access to the curriculum**

A number of participants spoke about their frustrations in not having access to a full curriculum comparable with hearing and sighted young people. A few participants with type 1 Usher, spoke about their frustrations of only skimming the surface of subjects or not being taught in the most appropriate manner. Linda and Faith commented:

It was good because I made a lot of friends, had a lot of independence. Had good fun. And I socialised. But in terms of education I wasn't very happy. I found it hard to follow and I missed a lot of stuff. At the time I wasn't particularly aware of it. But looking back it seems a lot more obvious now and I realise now that I should have been a bit tougher with them to make them more Usher aware. Whether with signing or different ways of educating that might be more suitable to me. But that was that time, I can't go back now. **(I)** (Linda, 46, type 1.)

I've got general grades, general levels. I didn't get any of the credit levels. As I was growing up and I went to deaf school, and they didn't teach us the full curriculum it seemed to be quite basic stuff. In reading things were really brief and summarised. When I was about 11 I started reading more things and became more aware. When I was 11, when I went to a new school, I did English and I was like oh my gosh what's all that stuff! The first time it was quite a shock. So my qualifications seem quite low. Secondary school was better. After school I went to college in (3.5 hrs away) and studied web development. And I got the qualification. that was an HND. I'm happy about that because I've got more equality, I'm on a par with everyone else. **(I)** (Faith, 23, type 1.)

Similarly Susan (48, type 1) said that it was only when she decided to teach herself to learn to read that she was able to succeed.

Other students spoke of being excluded from particular lessons due to health and safety concerns:

At my old school I wasn't allowed to do PE. They wouldn't really let me do drama, dance... I wasn't really do much in food technology. Because I'd struggle to see it and I'd knock things over. But I could cook I just needed to put things where I needed to put them, and not where they wanted them. So I never really used to do much. They used to normally get me to help someone else. I'd kind of work with someone else but they'd do it for me. Which was a bit rubbish. I do all of it now though. I'm allowed! (Jess, 15, type 3)

However Jess's mother was pleased that her new school was encouraging Jess to take part in activities, "they say come on Jess, stop being so lazy, get on with it. She's gradually just got used to doing them again. You know like with swimming she was scared." On the other hand Chris was angry that his teachers made him take part in sport even though he could not see the ball:

I had an up and down experience at school. Some teachers were good, some were awful, some wouldn't let me be excluded from things like cricket and hockey – I can't see the ball!! I would refuse to do PE at certain sessions and then I'd get detention for it. Things like that, I'd think I'm getting detention for my disability. That's not fair! (Chris, 33, type 3.)

Jess was keen to point out that people with Usher can still take part in lessons, but that they just might have to do things slightly differently. In response to a question regarding an imaginary situation where a pupil had been advised by a doctor not to study art, Jess said:

Not very fair. Because he could still do it but he could do it a bit different. Like he might do like more tactile pictures or make different stuff, not really draw it all the time. We do that at school. Quite a lot of them take art for GCSE. But we don't draw. If we can draw we draw, but most of us... I made like bowls, and painted them. I made a mosaic picture. I made a clay person that was fun. So we just make things instead of drawing. (Jess, 15, type 3)

Harry (15, type 2) however wished that he could avoid studying French because he finds the spoken language part too difficult. Nicola (23, type

2) looking back on her education wished that she too could have avoided studying French, whereas Richard (48, type 2) who had been excused from studying French, wished that he had had the opportunity.

Lastly a number of participants spoke about having to miss classes, especially in the younger years to attend speech therapy, which participants found once again singled them out from other students.

10.3.3 **Careers advice**

As discussed in chapter 1 on diagnosis, a diagnosis of Usher can often strike young people at a time in which they are negotiating the difficult years of adolescence. As well as impacting upon interactions with peers, as will be discussed in section 1.4, it can also affect the career choices that individuals make and the routes of study that they follow. Victor explains:

When I left school I was actually diagnosed as having Retinitis Pigmentosa at about 16 or 17. It threw me sideways. I didn't quite know... I tried to do A-levels but I couldn't really concentrate. I went to a college for Further Education. I didn't know what I wanted to do. (Victor, 56, type 3).

For Alex, resisting wearing his hearing aids until sixth form, due to a fear of looking different, meant that he felt that he did not achieve his full academic potential:

When I went into sixth form I got in-the-ear hearing aids, I thought that I've got to bite the bullet, but by then the damage had been done and I had lost interest in education by that point. I wandered for a few years through education. Then got into work through other means. (Alex, 29, type 3.)

This lack of direction, and difficulties in knowing what to do for the best was also discussed by other participants (in relation to career choices at school and later on in their careers). As mentioned elsewhere in this report, the uncertainty of the way in which Usher will progress can impact quite heavily on individual's lives; consequently some participants were unsure of whether to follow a particular preferred educational path even though it may be enjoyed for a limited time only:

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I was always, always artistic always doing design and stuff. So I did design technology, I did textiles, I did fine art. I didn't do fine art for GCSE I just went straight to A-level. So I knew at that point that's what I wanted to do, my whole life had been gearing up to this point, and I've always had in mind to do something in that area. Then I got told, at exactly at the wrong point, what was going on. To the point that the one real effect that I had, was that I couldn't really pick up a pen or a pencil for about a year. I just didn't want to face it. It was the one thing that really really affected me. The rest of my life I almost just completely carried on, but that was the one bit where I really felt, I don't know what I'm doing. Do I continue on with this stuff or do I find something else? In the end my art teacher sort of took me aside and said look, you know, you need to decide what you want to do. Gave me a pep talk really. And I realised well that the thing that makes me happy, so I want to do that for as long as I can. Get to the best point that I possibly can, and then when I can't do it anymore find something else. (Megan, 24, type 2).

Here we see that with support Megan was able to make her decision and move forward with her life and is now in fact successfully using her artistic skills in her chosen career. It is interesting to note that Megan's support does not come from a specialist teacher, but rather a teacher who had an interest in supporting her. Leah and Chloe's mother was also frustrated that careers advisors did not provide specialist support for her daughters and failed to understand the implications of a diagnosis of Usher:

Cos on the childcare courses you get placements, but once the placements find out what you are actually suffering from, they've got you on Health and Safety grounds. Straightaway! It was horrid. I mean the Connexions teacher at school knew what conditions they had, but pushed them to take different courses and stuff like that. But what are they going to do at the end of it?

For a lot of young people diagnosed with Usher whilst in education their condition had not impacted upon them, however others had to considerably alter their future career plans, Steven (36, type 2) for example had to give up a university place studying dentistry. A diagnosis of Usher meant that Rob was no longer able to follow his dreams of being a builder, which had a considerable effect on his wellbeing:

I used to be a builder. I was at college. I did two years when I was diagnosed as partially sighted. Health and Safety found out, kicked off, and escorted me off the site. And I won't allowed back. And I had no choice to give up that, and that was devastating for me because I'd worked so hard. I had put my heart and soul into that. And it was something I really enjoyed. And I could have seen myself owning my own little business. I had to give up that. I guess that's part of the reason why I had a breakdown as well. When I got diagnosed I couldn't get round Health and Safety. I begged, I pleaded, but it's a dangerous place isn't it? On a building site... so yeah I had to give up that... (Rob, 22, type 2.)

Likewise Lucy who received her diagnosis of Usher whilst at college had to give up her dreams of being a self-employed plumber as she was no longer able to drive nor to complete tasks safely. Fortunately however after a period of reflection Lucy re-trained as an audiologist – something that she suggests she would never have done had she not had Usher:

I think the deafness is part of me. Without it I wouldn't have become an audiologist. I've done my degree. I'm going to do my Masters. I was going to be a plumber and all that but I enjoy this so much more. I'm good at it. I think it's made me who I am. (Lucy, 27, type 2.)

Furthermore through her experiences of having Usher Lucy felt herself to possess greater skills and understanding when dealing with deafblind patients in clinics compared to her sighted and hearing colleagues. Richard and Ben also spoke about their determination to go to university in spite of their hearing loss (Richard's Usher was not diagnosed until he was in his 40s and Ben's biggest challenge was being a sign language user in a hearing world).

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10.4 Interactions with Peers

Interactions with peers are an important aspect of education and as well as contributing to an individual's sense of wellbeing can also impact upon educational outcomes. Participants who generally had good experiences at school tended to speak positively of their experiences, whereas those with negative experiences also tended to have negative experiences. For example Farah and Megan spoke positively of their experiences at school:

My friends help me. Lead the way at night. If it's sunny and I can't see. Before when there was no understanding of Ushers and I didn't tell anyone, I would fall over, and some people you know would get quite agitated with that. They'd be no support for me. No help really. But when you tell them that you have Ushers it's clearer and more helpful. **(I)** (Farah, 17, type 1.)

I've always been fairly independent. My teachers knew that. I told my close friends. I didn't tell everyone at school I didn't need to. Like I said nothing really changed. All the teachers were very supportive. (Megan, 24, type 2.)

Whereas participants who overall had more difficult experiences at school spoke negatively about their interactions with peers; Doreen and Susan spoke of their negative experiences:

Went to a school for the deaf. Boarding school. I was the only one that had Usher in the whole school... the other children used to tease me and bully me. **(I)** (Doreen, 53, type 1.)

I wasn't truly happy when I was at [residential school for deaf children] and I was trying to learn things, and I knew I had a vision problem without knowing what it was. I had to hold the book very close to my face, or go up very close to the blackboard, and people would hit me on the back of the head. I don't know whether it was other children or teachers. **(I)** (Susan, 48, type 1.)

Problems arose because of a fear of looking different and a desire not to be singled out, difficulties in communication because of the use of different methods, and specific bullying because of Usher focusing up on a (perceived) vulnerability and difference.

A number of participants tried to hide their deafness from their peers. Richard (48, type 2), Alex (29, type 3), and Vicky (44, type 2) spoke of refusing to wear their hearing aids. Likewise Callum (14, type 1) was refusing to allow his teachers to wear the radio transmitter he needed for his cochlear implant. For Callum this has had a significant impact on his learning, however as Vicky explains avoiding telling people about your hearing loss means often missing out on the small exchanges that contribute to getting on with others:

Secondary school especially... The usual teenage problems, the complexes people have but also I just didn't want to tell anyone that I had a hearing problem. I had no confidence about it. And I think that in itself caused a bit of a problem, I probably didn't pick things up, or people were joking around. (Vicky, 44, type 2.)

Vicky also highlights the fact that the teenage years may be difficult and people with Usher may also face difficulties with their peers for a host of reasons, not just because of their Usher.

Although the majority of young people did not want to be singled out because they had Usher, Harry's mother explained how the teachers had made all the pupils aware of his Usher:

They do assemblies don't they every year. For the new kids coming in. Just to highlight it, cos some of them are just so little and Harry's nearly 6ft, and you get the little year 7s that are coming in, are only like about this big, he doesn't see them, so they have to be aware that they have to look out for him really. (Harry's mother, 15, type 2.)

Likewise when Bethany (17, type 2) began her new college, she was anticipating that the college would tell the other students about her being deafblind, to save her the trouble of constantly repeating herself, especially since she felt her deafblindness would be difficult to hide as she has a guide dog.

Instead of hiding their hearing loss a couple of participants used humour to deflect any potential embarrassment. Ryan (17, type 2) and Lucy (27, type 2) explain their tactics:

Ryan: No. I take a joke with it to be honest. Cos like when I was born I used to get bullied. I just go along with it and laugh at it. Join in. So I say yeah, cool. I don't really care.

Mother: I think that you have more hassle because you wear glasses for short sightedness than you have because of the Ushers.

Ryan: Yeah. They don't really know about my Ushers. They just think I look like an idiot with glasses.

Mother: But then everybody gets that, whoever wears glasses.

Ryan I just go along with it. I just go yeah I know I look like a fool.

Mother: Yes I'm a young, sexy fool!!

Ryan: But yeah I just go along with it. But then it backfires on them and they just shut up. I think it's funny.

But I had a loop system that the teacher used to wear. And I used to wear. It was a big box in those days. But I made it cool, cos like the teachers used to leave it on, and go to the staff room, and I used to hear all the gossip. I was like, oh guess what they're all talking about in the staff room! (Lucy)

Difficulties in communication with peers meant that some participants were left feeling upset and detached from others. Dima commented:

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Yeah I like school but have not good to go to school because can't join in social. Sometimes I can't understand some people using their BSL, because my Usher. Sometimes I have to tell them repeat again, sometimes they feel annoyed to repeat it for me. It sometimes make me feel low. (Dima, 15, type 1.)

For Bethany there were difficulties because although her lessons were taught in spoken English, the majority of pupils used BSL outside of lessons. Since Bethany only had a limited knowledge of BSL, this made interactions with fellow students challenging:

...in lessons it's all oral, you have to use your voice because it's an oral school, so the lessons were fine, it was just the social side of it, and with it being a boarding school as well it was really difficult... It was really isolating. Very hard to fit in. (Bethany, 17, type 2.)

Lastly a number of participants spoke about specific bullying because of their hearing and visual impairments. Commenting on his experiences at both a residential school for deaf children and one for blind children Chris said:

but at both schools there was a lot of bullying. One you were bullied because you couldn't see, and then you were bullied cos you couldn't hear – so you couldn't win basically! (Chris, 33, type 3.)

For Olivia her experiences at a residential school for deaf children was also hard to listen to:

I was mocked, and “oh you’re stupid you can’t see”, and they would encourage me to go into things and trip over and have a laugh about it. So I was homesick and afraid of the dark, so I had to sleep next to the door, so I could see the light and I felt safe. And they would joke and close the door, and I would keep opening the door. And sometimes I would wake up in the morning maybe about 4 o’clock and I would be really nervous, I couldn’t go to the toilet, I was frightened. I had to wait. And I would just wait under the blankets, until 7am and the lights were on. And that happened sometimes every day, and that went on for 4 or 5 years which meant I was extremely tired. **(I)** (Olivia, 54, type 1).

Jess (15, type 3) and her mother talk about the particularly difficult time that she had at her residential school for deaf children:

Mother: Even though some of the others had Usher there, they could all see. And everybody thought because your eyes looked ok you could see as well.

Jess: They were just evil to me.

Mother: I know they were.

Jess: They were horrible.

Researcher: Was that the other children?

Jess: Yeah.

Such an experience led Jess’s mother to suggest, “the most difficult thing, was actually being at [school] was worse than going blind actually.”

Unfortunately for the participants who had left school these issues of bullying were not overcome whilst they were at school, however for students still at school the bullying had been resolved by moving school.

10.5 Discussion

10.5.1 Change

There are a number of features related to change in the stories people told about themselves. Some of these are change in the person who has Usher, because of Usher, but others are extraneous to that.

For individuals with Usher, change in their education was frequently related to other people's perceptions of change rather than their own perception of change in themselves. Changing schools for instance, a major event for most young people, was imposed on them because they had Usher, and not necessarily when they themselves saw need for change (e.g. Chris talked about the range of schools he attended). While in other fields (communication and mobility) people with Usher changed only when they felt they had to, in education others made this decision and often in the accounts above, it was not a good one for the young people (e.g. Faith.) As was often the case in other areas of their lives, young people with Usher at school and University did not want to look different (for example Michael explains how he pretends there is 'nothing wrong' with him.) Sometimes the changes were imposed in relation to what they might have felt was an identity which they did not want – for example using a different communication method, as Bethany found when going to a BSL using peer group, or Olivia, who did not feel comfortable using sign in school. Michael recognised himself as visually impaired only when attending a specialist college for visually impaired people.

Instead, in general, they would have preferred that the schools changed, and adapted for them rather than that they had had to change. In some cases, this did happen, for example, Abigail's teachers using the board properly but in other cases, the changes made were not helpful, for example where Faith was given large print but did not need it. Where changes were made, they did not always like it, particularly where they felt singled out by for instance, the use of a teaching assistant, as mentioned by Nicola, or by being with other people with disabilities, as mentioned by Jess.

Changes outside the individual have perhaps had more effect on people with Usher syndrome in the time span demonstrated by the project (up to more than 50 years since the oldest participant began school). Attitudes have changed as Olivia noted and more children attend mainstream schools, in a culture of more inclusion. Of the youngest age group about half attended a mainstream school whereas the oldest group, who knew they had Usher at school, all attended a special school. This reflects a change however not just in the attitude to schooling, but possibly in diagnosis of Usher for some at a younger age.

The effect of change in this way demonstrates that a change in the environment and the wider community can have at least as great an effect on the lives of people with Usher as can change within themselves.

10.5.2 **Predictability/uncertainty**

The uncertainty of what is in the future is also demonstrated in the accounts of education. Changing schools did not always happen at the right time for individuals; Chris's experience shows that he had to change schools at a moment which did not take account of his age and status in school, (in year 10).

But on the other hand, Michael recognised his visual impairment more when in the company of other visually impaired people.

Uncertainty about what is needed is also seen (second hand) in the attitudes of the education staff, who do not know what to expect, even specialist staff, who are perhaps trained in deafness but not visual impairment (as for example mentioned by Bethany and Chris). They might for instance prevent people from doing things which they would have enjoyed because they were unsure of the consequences, (Jess being excluded from cookery). On the positive side was Megan's teacher, who told her that she needed to think about what she wanted, which gave Megan the ability to see beyond her diagnosis, despite the uncertainty.

With a focus on education leading to careers, Usher has a particular effect on people choosing courses. Steven for example gave up ambition to be a dentist and Lucy to be a plumber. They cannot know

what they will be able to do when they leave school, or when they have achieved a degree, in relation to their vision. This can limit choices, as it did for Lucy, Steven and Rob. For others, as with Megan above, they made the decision to do what they wanted and only dealt with difficulties later.

The effect of the diagnosis of various types of Usher on individuals, particularly younger individuals where types were named through genetic testing, shows that choosing schools or educational support based on previous typology is unhelpful. It would not have been expected for example that a child with Usher 3 would need to attend a school for the blind and be learning braille, but Jess (suspected Usher 3) is, at 15, doing that. Even with the same type of Usher, and the same gene (presumably) the brothers Alex and Chris had very different school experiences. This demonstrates once again that individual decisions need to be made and that for those who are diagnosed with Usher, there is no outline pathway defined by Usher or by the type of Usher an individual has.

10.5.3 **Diversity**

Throughout the discussion of schools, further and higher education and the choices made, the diversity of choices made by individuals with Usher, their families, and education staff shows through. As outlined above, people with Usher followed courses in things which required vision (for instance, Megan) and changed their education to adapt to reducing vision. They studied in typical and large print, braille, spoken English, BSL and a combination of all of these. They followed courses in all main subjects, including PE and foreign languages and took a wide range of qualifications, depending on aptitude and interest.

For schools and education staff, the outcome of the interviews does not highlight a clear path to their role and responsibilities. Some students liked extra support and some did not. Some wanted to go to specialist schools and were happy there; others did not. Some enjoyed school and others did not. Some had good friends, who also supported them, others had no friends or were bullied. In all cases this is the same for all young people, or young people with disability. While there are clearly some areas for improvement, where diversity of experience is not of

benefit (as in teachers who listened to pupils and those who did not, and the concerning issue of bullying for some pupils) overall, it can be seen that asking pupils and their families what they want is likely to be of most benefit, as pigeonholing children due to deafness, to Usher, or to increasing visual impairment rarely seems to have had good outcomes.

11 Employment

Work plays an important part in self identity; competence at work is one area identified by Gullacksen et al (2011). It is also important for economic well being, and for the social benefits from engagement with society and making a contribution to the whole. From previous interviews with people who had Usher, many of them were not employed in paid work. 4 of 20 in Oleson and Jansbøl's (2005) study were in work in an ordinary workplace (though some were students and some retired). Gullacksen et al said that some of their participants had to stop work, or reduce their hours. 34% of visually impaired people interviewed by Douglas, Corcoran and Pavey (2009) in the UK were employed and 22% unemployed.

Consequently this chapter will begin by exploring the employment experiences of the participants in this research project. We shall then move on to look at the challenges that participants faced in relation to employment, specifically in accessing employment and when in work the types of jobs that people with Usher do. The third section looks at the support that people in employment received. The final section brings together the themes of change, predictability/uncertainty, and diversity to examine what is special about the Usher experience of employment. .

11.1 Overview

Participants in the research project were employed in a number of professions including as an occupational therapist, independent financial advisor, running their own businesses, BSL tutor, audiologist, office administrators, communicator guide, and jewellery designer. Using the responses from the 32 adults in the project we see that 8 (25%) of participants were working full time and 9 (28%) were unemployed. Overall 17 (53%) of people were involved in some kind of employment. 4 (13%) participants were full time mothers. One person (3%) was in further education and one person (3%) was unable to work due to illness.

Although the sample is small, people who had type 1 Usher, and who in the case of adults were all BSL or SSE users, were less likely to be in

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fulltime employment and more likely to be unemployed or working part time than people with type 2 Usher. 1 person (8%) with type 1 Usher was working fulltime, whereas 4 people (25%) with type 2 Usher were working full time. Similarly 5 people (42%) of people with type 1 Usher were unemployed, compared with 3 people (19%) of people with type 2 Usher. Half of the people with type 1 Usher were working part-time compared with 3 (19%) of people with type 2 Usher

	Type 1 (n12)	Type 2 (n16)	Type 3 (n4)	Total (n32)
Work full time	1 (8%)	4 (25%)	3 (75%)	8 (25%)
Unemployed	5 (42%)	3 (19%)	1 (25%)	9 (28%)
Work part-time	6 (50%)	3 (19%)	0 (0%)	9 (28%)
Child care	0 (0%)	4 (25%)	0 (0%)	4 (13%)
Further Education	0 (0%)	1 (3%)	0 (0%)	1 (3%)
Unable to work due to illness	0 (0%)	1 (3%)	0 (0%)	1 (3%)

This appears to be slightly different to the balance for participants in Oleson and Jansbøl's project, although the numbers in both are small and the social conditions in the UK and the Nordic countries are different. In their study, more deaf people than hard of hearing people (their terms) are in work.

The experiences of participants in the younger and older adult age groups did not differ significantly. For example 4 (29%) of young adults were working full time compared with 5 (28%) of the older adults. 4 (29%) of the younger adults were unemployed, and 4 (22%) of the older adults were unemployed. Similarly 4 (29%) of younger people were working part-time as were 5 (28%) of the older people.

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	Younger adult (n14)	Older adult (n18)	Total (n32)
Work full time	4 (29%)	5 (28%)	9 (28%)
Unemployed	4 (29%)	4 (22%)	8 (25%)
Work part-time	4 (29%)	5 (28%)	9 (28%)
Child care	1(7%)	3 (17%)	4 (13%)
Further Education	1 (7%)	0 (0%)	1 (3%)
Unable to work due to illness	0 (0%)	1 (0%)	1 (3%)

It is worth noting however that for the people involved in part-time work some were happy to be working reduced hours as it fitted in with their family life, and meant that the fatigue associated with Usher was more contained. However other participants were looking for full time employment. Similarly some of the participants were happy to be unemployed and were not seeking employment, whereas others were looking for work.

Due to the nature of employment this chapter will focus on the experiences of the younger and older adult groups. However it is worth noting that the young people in the research project were thinking about careers in teaching, brick laying, fashion design, computing, and as a professional athlete. Additionally a couple of the young people spoke about the difficulties of getting part time jobs which are often seen as an integral part of the transition to adulthood. Farah (17, type 1) suggested that she would like to have a part time job but she did not think it would be possible because she would need an interpreter to be with her at all times. Bethany (17, type 2) too was keen to get a Saturday job, for the social aspects as well as something to keep her busy, however she was finding it difficult to get work. Although she had been in contact with her social worker to ask for his assistance in helping her find employment,

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she was frustrated with his lack of progress. Dima (15, type 1) however was not keen on getting a part time job and Harry (15, type 1) was too busy competing in sports to do so. Rosie (21, type 2) looking back on her experiences of work as a teenager, explained that due to the long days travelling to a school with a resource centre far away, and the extra effort involved in keeping up with schoolwork regrettably she was not able to take a part time job.

11.2 Employment Challenges

11.2.1 Accessing Employment

Challenges to accessing employment arose at two distinct points in the lives of participants. For participants who had knowledge of Usher whilst at school there was the issue of accessing employment in the first place. Secondly a number of adult participants had to deal with the impact of making changes to their current working practices when they were diagnosed with Usher or when the progression of their Usher was such that they could no longer continue in their current job in the same way.

As has been shown elsewhere in this report, participants had a desire to get on with their lives regardless of Usher, yet at the same time acknowledged some of its limitations. Participants recognised that there were some jobs that were not possible as Steven explains:

It's really difficult. I wish that I could turn round and say pursue whatever you'd like, pursue your dreams sort of thing, but at the end of the day when it comes to career you've got to be quite realistic. (Steven, 36, type 2.)

Likewise Alex (29, type 3) knew that he could not be a taxi driver, and Dave (46, type 3) cautioned, "You're never going to be an airforce pilot, you're never going to be a driver." Yet at the same time Dave said, "I would say never let it stop you doing anything. If someone puts a brick wall in front of you, you've got to get over it. Whether you get round it or under it." At first glance this may seem to be a contrary position to hold, but Dave, like other participants was reiterating the belief that a person with Usher should follow their dreams by focusing on what they can do, rather than what they cannot.

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As was discussed in the chapter on education, one of the challenges for young people with Usher was planning for the future. The knowledge that she had Usher and having been told, “you’ll have to stop work by the time that you are 30” **(I)** meant that Faith (23, type 1) “Never thought more than short term, just a few short years.” **(I)** Consequently it had taken a Faith a few years to believe that she could get the qualifications that she wanted and to be employed in a job that she enjoyed. Likewise Chris questioned the futility of studying if he was not going to be able to use his skills in the future,

I used to say if my sight gets worse I won’t be able to [do my job]... I used to think I’m going to do all these degrees and things to get there, am I wasting my time? (Chris, 33, type 3.)

In order to deal with the uncertainty regarding the way in which Usher develops, a number of the young adults, looking back on their late teens and early twenties, decided to stop worrying and just focus on the present. For example Megan (24, type 2) in choosing to carry on with the career path she wanted to follow explained, “I just decided that you can’t predict the future, you can’t say for certain what’s going to happen.” Similarly Emily said:

Because for years and years and years I’ve been asking questions about my future. What can I do? What jobs am I going to get? How’s it going to get worse? And I kept asking myself those questions and then in the end I’ve just given up. Do you know what? I’m not going to ask myself any more questions about the future because at the end of the day we don’t know and we have no idea about how bad by Usher is going to get. It could stay like this, it could get worse, so I do think it will affect my career, because what I want to be in the future I’m not sure if I’m going to be able to achieve that because of the Ushers. **(I)** (Emily, 27, type 1.)

Participants who had been working, but then because of changes with their Usher had to give up, also faced challenges. As has already been discussed, these crises can be as upsetting and as difficult to deal with as the initial diagnosis of Usher. Michael, who had previously been employed in a number of roles, found that it was no longer possible to

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continue working and chose to resign rather than getting 'sacked'. He describes how it left him feeling and the support that he got from his GP:

I think that knocked my confidence quite a lot. I just stayed in my room for 5 years then. I didn't go out or get a job or nothing. I was on Job Seekers but they told me there was nothing they could do for me. Go down the doctors, get a doctor's note and you never have to go back again, and just left me in my room. Just didn't want to help me at all. (Michael, 34, type 2.)

Similarly Carolyn (38, type 2) who had been working at the time of her diagnosis was signed off sick by her doctor:

When doctors found out and I enquired a bit more. He said I'm going to give you sick note for the next six months. He just said I couldn't work any more. So that's when I decided that I'd get married and have children, to keep my time occupied. (Carolyn, 38, type 2.)

Alice (41, type 2), Doreen (53, type 1) and Nicola (23, type 2) also spoke of how 'straight forward' it was to be signed off sick from work after an initial diagnosis of Usher.

Linda (46, type 1) had had to take medical retirement from her job because,

"My eyes had got worse over time and I was getting more tired. I was finding it more stressful with the travelling, particularly at night." (I)

However after time to reflect and recover Linda had sought part time employment elsewhere. Linda had only recently taken up this job and was wary about it being 'too much' however she was pleased to be back in employment. These uncertainties about Usher, for example the effect of stress on the eyes, are something else that participants have to contend with. Victor (58, type 3) too had had to take medical retirement after 25 years in the Civil Service. Although still quite angry and frustrated with his situation he was very active in with his hobbies and voluntary activities. It must be remembered that lack of paid employment does not mean that one is not actively contributing to society.

Part time employment or reduced hours was the answer for some people to enable people to keep accessing employment. Reduced hours

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were useful in two ways – one to prevent tiredness and ensure that participants could continue with their work, and also as in Vicky's (44, type 2) case, it meant that she arrived and left work in daylight hours.

Returning to Michael's experience, after 5 years with the help of a deafblind worker from social services, he was able to attend a college for visually impaired people. At the college he retrained, learned rehabilitation and mobility skills, and began a six months' work experience course which eventually led to 16 hours employment a week, which he was very pleased with.

The importance of retraining in order to access employment was reiterated by other participants. Emily (27, type 1), Lucy, (27, type 2) and Megan (24, type 2) were satisfied in their current employment and said that they would 'just retrain' if and when it became necessary. It is interesting to note that all three of these women had left university recently and perhaps saw themselves as having a greater set of skills and opportunities available to them. Participants who had had time off to care for children, felt even more distanced from the labour market and were concerned about the jobs that they were trained for would no longer be possible. Nicola and Julie shared their experiences:

I'm fully first aid trained. I've got an NVQ in bar work. Serving. I went to catering college. But I couldn't do anything that I'm trained to do. I'd have to find completely something else. Something that involves, not moving, good lighting, things like that, cos it would be hard going back behind a bar. It would be impossible. (Nicola, 23, type 2).

The problem I've got is what I trained to I can't do (nursing). So I've got no qualifications in what I've got to do, which is in an office or something like that. I'm going to find it really difficult. That's another thing as well trying to get a job to fit round (children) and the fact that I've got no qualifications and that I've got Ushers as well, I'm probably way down on the list. You know I am going to find it hard. (Julie, 38, type 2.)

Although still in employment, Richard shared Julie's worry that an employer would not want to employ a deafblind person:

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There's two things really – I know I've only got a limited amount of work time left, and I need to up the income that I am earning, but to counter that there is a concern in the back of my mind, who'd want to employ someone who's losing their sight and is half deaf. There is that issue as well and that has always been an insecurity of mine if you like. (Richard, 48, type 2.)

Richard also emphasises the point that although still working he had to contend with the fact that his Usher could change at any moment, and as a father to two small children he was concerned about working long enough to support his family. These concerns were also shared by Steven (36, type 2) who also had a young family.

Participants were however keen to point out that difficulties accessing employment affected everyone – not just those with Usher. Ben (30, type 1) spoke of the global credit crunch, Dave (46, type 3) bemoaned immigration from Eastern Europe, Hamid (44, type 2) had to find employment that fitted in with his caring duties, and Lucy (27, type 2) spoke about cuts to the NHS.

Lastly, echoing the poor experiences of young people in accessing careers advice, only one person spoke of dedicated services to help them into employment. Jayne (28, type 1) was keen to recommend the services of a Disabled Person's advisor at the Job Centre, who she felt was crucial in understanding her situation and getting her into employment.

11.2.2 Types of jobs that people do

Despite the belief held by many participants that a person with Usher could do whatever they wanted, in reality participants were often working in specific settings. Of the 18 people who were working (either full-time or part-time) half (nine) were working in organisations that provided services and information for people with disabilities, particularly those with sensory impairment (this was reflected also by Gullacksen et al, where half of those who were working were employed “in relation to others with a sight or hearing disability” (book 5 pg 24). A further 4 were employed by the NHS. Of the 9 unemployed people 3 were looking, or would like to work for an organisation that has a specialism in hearing

and/or visual impairments. Of the 7 BSL/SSE users in employment (part or full time) six were employed in these specialist organisations.

Participants wanted to work in such organisations as they felt that they had the appropriate understanding and had made adaptations or were amenable to change, particularly with regards to communication as Jayne explained:

I find it more comfortable working with people with disabilities, because there's more understanding. A bit more patient. Like they might expect you to be quicker, or like if your English isn't very good. I find it more comfortable working somewhere like this. People are a lot more patient. **(I)** (Jayne, 28, type 1.)

Similarly Abigail (29, type 1) commented, "I would like to work in Sense – that would be much easier for me. Admin maybe." **(I)**

Nevertheless spoken English users also suggested that working within d/Deaf organisations was a solution to overcoming communication issues. The sense of relief experienced by Hamid in working in such an environment is evident:

I then got an admin job, I was studying there, I was working there. I could communicate with the students. I felt comfortable there. That was like, 'Ah this is it. This is the solution. (Hamid, 44, type 2.)

Victor (56, type 3), also a spoken English user, suggested that young people with Usher would be better off working in the charity sector or office based work. Likewise Steven who was struggling in his high-powered job suggested that it may be better to work in a more 'forgiving' environment:

Sort of job I do. I'm not working for a charity or anything. It is a commercial environment. There is a lot of pressure on what we do. We have to get things done, and on time. You don't want to appear like the weakest link. (Steven, 36, type 2.)

As well as being understanding and providing support in relation to communication, participants suggested that such organisations were understanding about blindness and Usher. Important for Faith was the fact that "I'm just a member of staff" **(I)**:

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It's great here obviously because of their awareness. There's some staff that are completely blind, there's me with Ushers, and people understand, and I'm not the only one now. It isn't as much of an issue. It's not a problem. We all understand each other. The staff are natural. I'm just a member of staff. I don't have to say anything. And everyone's aware of disability so it's much better. **(I)** (Faith, 23, type 1.)

Emily's employers were particularly aware of her changing needs as a person with Usher:

They understand that I can't work at night time. I'm not able to support my service users at night time. They're also very aware because I do work for a deafblind charity, I'm quite lucky because they have the awareness already. They do support me and they have actually asked me to provide a hospital report, next time I go to the hospital, so they can assess what I can do long term obviously because of my Ushers it can be quite risky, trying to support clients in the community, but they do want to watch my Ushers, its progress, and make sure it's not deteriorating. **(I)** (Emily, 27, type 1.)

As well as working in organisations which support people with sensory impairments participants suggested that it would be better if they did office work or as Vicky (44, type 2) suggested, "something that doesn't involve moving around too much." However a couple of the young male adults, along with teenagers Ryan (17, type 2), Callum (14, type 1), and Daniel (15, type 2), resisted this type of work:

I don't want to be stuck in some office. I think that it'd drive me crazy – it really would. I'm more of a hands on person where I like to be active, do things. It's hard to find a job doing that. (Rob, 22, type 2.)

Chris spoke about his determination, combined with his bloody-mindedness, which meant that he succeed in his job in the health service:

I know that these types of jobs are probably more suited for me because I can sit on my arse and tap on the computer, whereas really I want to do something completely different, but I may not get there. So there's always that... you as an individual, and that

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arrogance in you that sometimes you don't always help yourself if you like, you make it more difficult for yourself. (Chris, 33, type 3.)

Self-employment was suggested by a few of the older participants as a way to continue working, as participants could have more control over their environment, working hours, and not have to explain themselves to others. Dave and Olivia commented:

The shop is laid out exactly the way I want it. I can tell you where everything is. All the boxes are numbered on the inside. Everything's laid out strategically. You noticed the yellow door? (Dave, 46, type 3.)

I'm happy working here. It's very easy for me. It's right by my home, which is less stress, so I don't have to worry about being driven. It's perfect. I enjoy the work. **(I)** (Olivia, 54, type 1.)

Nevertheless Dave (46, type 3) complained about the lighting in his shop, which he was unable to change because he was only renting, whereas Olivia (54, type 1) had had a purpose built teaching space constructed to her own specifications. Self-employment also allowed participants to spend more time with their family. The 3 participants who were self-employed fell into the older adult age group which fits with national trends on self-employment (ONS, 2013).

Finally it must be recognised that participants with Usher bring useful skills and knowledge to the workplace, specifically because of their Usher. Vicky (44, type 2) suggested that she was more understanding of patients' needs and a better communicator because of her Usher, for example she picked up on small changes in body language and looked directly at people when speaking to them. Lucy who was an audiologist commented:

I'm good. It's not big headed but I'm good at my job. I have a much greater understanding than other professionals. When I was doing my training and this woman was fitting a man, totally blind, I just had to take over I couldn't watch. She was said something like, can you understand what I am saying now you can't lip read me, and I was like he couldn't lip read you anyway! (Lucy, 27, type 2.)

Rob's experiences had also led him to want to train to become a counsellor:

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I wanted to help people come to terms with Charles Bonnet syndrome and that. So that they've got somebody to talk to without being judged. Cos as a sufferer I know what it's like and I know how scary it can be. I also lived with the fact that I thought that I was mad. So who better, someone who knows, someone who understands. (Rob, 22, type 2.)

11.3 Support for Employment

The majority of participants in the project who were working said that they received support in employment. Generally participants spoke about support in broad terms whereas a few specifically referred to Access to Work. Access to Work is the Government scheme which “assists disabled people who are in paid employment, or with Job/Work Trial, by providing practical support in overcoming work related obstacles resulting from their disability” (Department of Work and Pensions.)

The support that participants received generally fell into three categories – equipment, transport, and communication. The most common form of support that participants received was in relation to equipment, for example telephone amplifiers, high contrast keyboards, task lights, and antiglare windows. Chris (33, type 3) was also provided with specialist software on his laptop and Lucy (27, type 2) was hoping to be able to get specialist equipment with brighter lights on it.

Some participants received support in the form of transport in getting to and from work. Steven (36, type 2) for example travelled to and from work by taxi every day. A couple of participants when asked if they received support for transport were keen to point out that they did not because they did not need that level of support. Faith (23, type 1) commented,

“I have access to work, but I don't need a taxi. I can see. I wouldn't like that. I'd feel sly. I can get the bus fine.” **(I)**

Michael (34, type 2) used the bus to get to work, he had been contemplating applying for Access to Work, however he felt that it was 'too much bother.' Lucy too was sometimes let down by the

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administrative side of Access to Work, and although support may be available in theory, in practice it does not always work so well:

It was a bit difficult when I was living in a little village before, bus used to be once an hour. So I used to do the whole Access to Work taxi thing. That used to drive me crazy, because the taxi would be late or... and you end up being really poor because I was paying for it out of my own pocket and then claiming some of it back through Access to Work but it would come ages later, so things like that weren't so good. (Lucy, 27, type 2.)

Participants also received support around communication. For some participants such as Faith (23, type 1) and Emily (27, type 1) who were in organisations that employed other Deaf people the support was less formalised and on more of an as-and-when needed basis. Whereas Jayne (28, type 1) working in a setting with other people with disabilities, but no one else who was Deaf, had interpreter support for key meetings, Hamid (44, type 2) a spoken English user working in a d/Deaf environment preferred using the in-house CSW (communication support worker). Of all the participants only Ben (30, type 1) and Chris (33, type 3) had full time communication support. Ben had a BSL interpreter and Chris had a lipspeaker. Without this support participants would not be able to continue in their employment as Chris explains:

I have a fulltime lipspeaker through Access to Work. I told Access to Work that I can't treat patients unless I have a communication support worker fulltime. My manager is quite good in saying, yeah you have to have a communication support worker... I've fulltime support basically. (Chris, 33, type 3.)

Despite the statutory requirement to provide people with the appropriate support, a couple of participants spoke about the difficulties that this entails. As Lucy (27, type 2) was working and studying at the same time, Access to Work and the Disabled Students' Allowance were in dispute over whose job it was to provide specialist equipment. Although speaking from around 15 years ago Julie found that her employer only did the bare minimum to support her:

They were helpful to begin with, but I feel as though it were because they had to be. They went down all the right channels of you know,

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having the meetings and things like that. It was a bit distant. I felt that they could have done more but they didn't. They just let me go. (Julie, 38, type 2.)

Chris too felt that he had to fight to get the support that he needed:

My employers are reasonably good and supportive, not without me having to shout, and running to my managers, taking things to HR, and looking up on the Equality Act, knowing my rights. Yeah they do support me, but I do find that I have to push, if you like have a strong stance on why they should, you could say it's almost a form of indirect discrimination under this law, then they change things because they don't want to look like they're not meeting your needs. (Chris, 33, type 3.)

Chris felt further frustration because of the review process of Access to Work and cuts to funding:

Access to Work are rubbish. Absolutely! They fund you, they review you in 2 years, and all they want to do is cut what they fund you. You have to argue out your case, why you should keep the budget as it is. (Chris, 33, type 3.)

Vicky (44, type 2) also suggested that it was because she was a long standing member of staff, had good relations with her boss and "they respect me as a therapist" that adjustments had been made, maybe suggesting that otherwise adjustments would not have been provided. Alternatively perhaps participants do not recognise their entitlement to support regardless of length of service or personal characteristics. Furthermore participants need to know what support is available before they can make use of it. Alex (29, type 3) explained, "I don't use any (support) because again I don't really know what I can use to help me really, it's a bit difficult."

Ben's (30, type 1) experiences highlight the differences between the daily communication support that was a positive feature in enabling him to go about his work, and the attitudes and discrimination that he experienced in not being able to attend training courses or invited to meetings, leaving Ben to suggest, "I think they've just got it into their

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head, oh Ben, you've got Ushers you can't do it." **(I)** Ben continues further:

... don't patronise people who are Deaf with Ushers. Don't do that. That's the main thing. Be equal. Be equal to hearing people, equal to people who are just deaf. It's all about equality please... they thought that I wouldn't be capable of doing my work. I wasn't included in some of the meetings that I could have gone to. They kept me to one side. I don't know whether they were concerned that I might have had an accident, but treat others with Ushers as an equal... I don't know whether it's naivety, discrimination, or patronising. **(I)** (Ben, 30, type 1.)

Vicky's comments on the good relations with her employers, highlights the importance of emotional support and understanding from employers. As seen above, participants tended to gravitate to organisations that work closely with people with disabilities/sensory loss as they felt these groups to be more understanding, as Faith explained:

My boss is great. They all learned to sign. There's a relaxed atmosphere. It's better than my old job at ASDA. They didn't understand about being deaf, they had no understanding. Not good. **(I)** (Faith, 23, type 1.)

Although not part of the formal support offered to him, Michael (34, type 2) appreciated the fact that colleagues were able to support him at social events such as Christmas parties. Likewise Lucy (27, type 2) mentioned that she had no shortage of willing volunteers to look after her guide dog whilst in clinics with someone who was allergic to dogs.

Alex also highlighted the fact that because of the way in which participants experience their Usher they might only need support in particular lighting conditions, or at particularly times of the year, this may mean that employers 'forget' that an individual has particular needs, as he explains:

There's about 300 colleagues that I work with. Not all of them know that I've got sort of a visual impairment, and even my line managers and stuff like that they forget I've got a visual impairment... And that's why I call it a hidden sort of disability, they sort of say to me oh yeah I

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forget that you can't see like at the sides, and they turn away from me. (Alex, 29, type 3.)

In accessing support participants need to make their needs known. As seen in the chapter on mobility, issues with mobility are harder to 'hide' if one has a cane or a guide dog, or alternatively make it easier because an individual does not have to state their situation explicitly. Participants who dealt with the general public and had high levels of support, said their clients were not bothered by the situation, and often did not seem to notice. Also as mentioned throughout this project participants were keen to tell other people with Usher that letting others know about your Usher is beneficial in the long run. For most participants this was straightforward, however some participants rejected having a label put on them, as Julie (38, type 2) commented,

My husband said the other day, you could do a job and have a badge that said 'I'm partially sighted' but I said yes but it's not very nice having a label.

In particular Steven was reluctant to let his needs be known as he did not want to be viewed as 'the weakest link':

(Colleagues) know that I have got a condition. They know that it's Usher syndrome, but I don't tell them how bad it is at the moment. So I don't mention to them that I can only read with one eye... it's not going to inspire confidence if you start telling people that you have trouble reading. So I don't mention it. (Steven, 36, type 2.)

Likewise in meetings Steven could not follow what was being said but chose not to 'hinder' the meeting by asking people to raise their hand when speaking, and instead afterwards,

I just follow up with someone else. I didn't quite catch what so and so said about... something along those lines, sort of thing. That clears things up. That's how I do it really.

Consequently life for Steven was very stressful:

It gets frustrating. You get angry. You do find yourself having to explain yourself all the time. You get fed up with it. Hence at times I'd rather be on my own. Left alone sort of thing. I'm the only person that

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I'm accountable to. It's trying to measure up to people's expectations of what would happen if I was normal. But I'm not. So life for me is a lot, lot harder, than a normally functioning person. (Steven, 36, type 2.)

It seems however that Steven was reaching a turning point, as identified elsewhere that participants will continue in a particular manner until they can do so no more, saying in his interview "I am interested in knowing if there is anyone else like me out there. Working in a commercial environment. Full time job."

11.4 Discussion

11.4.1 Change

Work revolves around change for most people – they move from education into work, they change jobs, leave a job to raise a family, eventually they leave work for retirement. It is not unusual then that all of these things happened for people with Usher but there were some differences. For example, some had to revise career or job planning because of Usher. Steven was unable to study dentistry, Julie could not go on nursing, and Brian could not find a job as a gardener. Others worried that they would have to change jobs in the future; like Michael, retraining after 5 years of unemployment. Although many people no longer consider that they are likely to have jobs for life, this is particularly so in the case of Usher, where participants were unsure how long they would be able to go on doing their jobs (for instance Richard, talking about how he believed he had only a short time to go on working).

There was a change also in other people. Julie expressed her frustration that while initially people had wanted to help her, this quickly drifted away. Michael however found that people were prepared to learn to support him.

For the young people, some expressed a desire not to work in an office (Ryan, Callum and Daniel). They wanted to do practical work. The experiences of older people suggest that this may not be the case when they get older. Vicky suggested that office work might be the only kind of things she could do, and Abigail suggested that she would like that

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kind of work with an understanding organisation. These young people had not perhaps yet had much experience of the changes Usher can bring and the compromises they might have to make. However having said that, Megan clearly became determined **not** to give up her dream of working in arts; she took up work which requires fine vision, for as long as she can. She has decided **not** to make a change until she had no choice

11.4.2 **Predictability/uncertainty**

The participants (at least the slightly older ones) knew that they had limited options. They knew they would not drive or fly aeroplanes. However, there were many things about their lives in employment that were not so certain. Julie felt forced out when work failed to support her adequately, and Carolyn was declared unfit for work by her doctor, perhaps to her surprise. Chris was unsure if he was wasting his time with his training, because he would not be able to do the work he had trained for. These decisions were beyond their own control.

They also had to change the way in which they accessed jobs. Ben and Chris both had full time support, one from a lip speaker and one from an interpreter. But it was not clear to them, or to others, how long support would go on –or they had to fight for it. This unpredictability - not in the participants themselves but in how and when they would get support - appeared to have a significant effect on their self perception. It might not be that **they** decided they could not work any more, but others decided for them.

Similarly unpredictable was how hard work might be for them. Linda talked about how tired she was, because of her deteriorating eye sight. Several had to work part time because they needed to travel in daylight hours, or because work was unusually tiring (they had to concentrate so hard to see and hear). Their colleagues might or might not know that they had difficulties with vision (or hearing) and their colleagues' responses were unpredictable.

A number of people found the certainty they needed about employment within organisations for people with disabilities (especially those with sensory impairment). They felt that in these situations they could expect

to find understanding and that, particularly for those who used BSL, their communication needs would be met.

Nevertheless, some decided to continue to work alongside the unpredictability of commercial sector jobs where they were not sure how long they could continue. This might be because they wanted to - Megan's work in jewellery, or because they were providing for a family - Steven in a high powered office job, but in these situations they were living with the uncertainty and unpredictability of future employment.

11.4.3 **Diversity**

As mentioned above, people with Usher who participated in our survey had a wide range of employment. They worked in administration, creative arts, health service professions, helping professions and in finance. Our web research showed people with Usher working as a journalist, a punk musician, a speed skater and more. Some had chosen to take time from work to bring up families. It is true that others were not able to find work and that they found this difficult. In many cases this was at least partly, and often probably mainly, because of their vision and hearing difficulties.

It is true that they also had options which were put aside because of Usher, - as well as mention of driving and flying aeroplanes, real plans for bricklaying and plumbing were shelved. Others were not able to continue with the careers they had wanted. While having a family was a choice for some, for others it was a pragmatic decision made when work became impossible - such as for Carolyn, who was signed off by her doctor. But some made new careers, found work in new sectors, and developed businesses which suited them. While Usher did certainly limit them, it did not stop everyone completely.

The participants also found and used a range of ways to make things work, being very creative with adaptations. Some worked part time to overcome fatigue, others developed their own business to manage their time and environment, some changed their travel arrangements - working only in daylight or getting support for a taxi. Some had specialist support at work, such as interpreters, while others found ways to manage themselves and their colleagues, more or less successfully. Others found such support within organisations for people with sensory

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impairment, where there was more understanding. They had particular skills, not just in finance, health care and so on but in understanding the demands of life – as mentioned by Rob and Lucy in relation to their ability to do their jobs and to help others.

Employment was a difficult area for some, which showed up problems and changes in relation to Usher. Some participants were not able to overcome obstacles to working but others met this challenge in a variety of creative ways. The younger people have still to move into work and suggested a variety of interests which might lead to careers. It is hoped that they are able to fulfil them.

12 Leisure Activities

This chapter offers an exploration of the leisure activities that people with Usher are involved in and tells us more about the lives and experiences of people with Usher. First we look at the leisure activities that the participants in the research project engaged in, investigating both individuals' desire and determination to do things whilst they 'still can', as well as the leisure activities that participants are prevented from taking part in because of Usher. However a large majority of people involved in the research project continue to take part in leisure activities by making adaptations and adjusting to new ways of doing things. A closer look at these adjustments and adaptations will make up the second section of this chapter. We shall then move to explore groups that specifically cater for d/Deaf people, blind people, and people with Usher, and suggest that participants who faced the greatest challenges in communication and mobility were particularly drawn to these groups. We shall then draw together the findings of this section around a discussion of the themes of change, predictability/uncertainty, and diversity, which together help us understand what is unique and distinctive about Usher.

12.1 Taking part in leisure activities

The participants in the research project were involved in a variety of leisure activities including gardening, snooker, attending church, bingo, tai-chi, Slimming World, raising awareness of Usher syndrome, watching TV, baking, keeping budgerigars, swimming, drinking, going to the gym, foreign travel, yoga, gaming, football, and going out for meals with friends. There were a small number of parents who wryly mentioned that nowadays all of their free time was taken up with child care!

In exploring the theme of leisure activities we can see some of the inspiring successes of people with Usher syndrome - brothers Chris (33, type 3) and Alex (29, type 3) both played VI cricket for England, Harry (15, type 2) was training for the Paralympics, Kat (17, type 1) was competing for Team GB in her chosen sport, Victor (56, type 3) had composed and performed music that had received international acclaim,

a couple of people had been Sense's Deafblind Person of the Year, Victor and Susan (48, type 1) had met with Members of Parliament to lobby for the needs of deafblind people, Rosie (21, type 2) was planning on trekking the Great Wall of China for RNIB, and Bethany (17, type 2) had raised over £40,000 for Sense.

Regardless of whether it be training for the Paralympics or going for a walk in the park, composing concertos or listening to music in your bedroom, travelling the world or visiting your local pub, what is important for people's well-being is to be engaged in the activities that they find rewarding and fulfilling . Not everyone can be a Paralympic athlete and not everyone would want to be; what is important is for individuals to be able to spend time doing the activities that they enjoy. In their own words here are some examples of the leisure activities that participants were involved in:

Well I love football. I do some sports. Maybe play football or tennis, or do some jogging outside. I also watch quite a lot of TV. I like to surf the net on my computer quite a lot as well. I'm starting to read some books. (Hasan, 15, type 1.)

Partying! When I finish work I go home, and I'm either asleep or partying. **(I)** (Jayne, 28, type 1.)

I do like socialising. I like eating out quite a lot. I do enjoy eating out... I like my cricket. Play blind cricket. I did the Black Mountain challenge, and do like events that the college holds. I do all sorts of things really. Keeps me busy. (Michael, 34, type 2.)

Leisure activities are one way in which individuals can build self-esteem, this was particularly so after the initial shock of diagnosis. Engaging in leisure activities is one way to boost self-confidence and to demonstrate to others that an individual is more than just a person with Usher. The boost to Ryan's self-esteem in playing for his local pub team, despite his Usher, is evident:

Every people that I've played against, they said I was really good. They said I just need to take my time. And obviously my mum's usually there (mum works in the pub) with me and she says thank you for saying that. And they say why did you say that and she says

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because he's got tunnel vision. And obviously mum talks about it. And they say I'm a really great player. I just need to take my time. I'm pretty chuffed with myself but still nervous. Cos like if I fail a shot or something. I just think that I'd take it out on myself, and my eyesight. So everyone says I'm a good player... So people were saying I'm a good player and just need to take my time, plus I've got tunnel vision, and can't really see. Look around. And when I look down it's all blurry. So it's even twice as hard. (Ryan, 17, type 2.)

Likewise Richard's pride in taking part in hikes with his friends, despite his worries about his vision, is clear:

We did a 3, 4 hour walk in Pennines... With my sight what's it going to be like? And it was tough, but it didn't stop me what I was doing. I was determined to go and enjoy it. It was fantastic. It was hard. It was horrible... It was hard going, but you're very much aware that you're climbing up and down things, and I'm thinking my vision isn't as good as everybody else's. But you know what I can do it! My friends actually said to me after, we wondered what you'd do. But you only get once chance in life to do things, and I know it's a bit dramatic, but if I was there tomorrow I'd do it again. But I really enjoyed it – at the end of it! (Richard, 48, type 2.)

12.1.1 **Doing the things that you can whilst you still can**

A number of participants spoke of a desire to do and see as much as possible whilst they felt they were 'still able':

I would like to travel more in the world. See as much as I can. See, hear, what I can. Before I get to the stage where I'm not able to. (Alice, 41, type 2.)

...while you still can do things like go rock climbing, or do certain activities do it now while you still can. See the world while you still can. Try and do things while you're relatively independent. Enjoy life. Then worry about it when it comes. When you start to find that your condition starts to affect your mobility... living day to day life and stuff. But at least you've got your memories and at least you've done what you've done. (Steven, 36, type 2.)

I know that I'll end up losing my vision completely, one day, but you know I'm not scared. I'm not scared... how can I put it... watch as much porn as you can before you lose your sight! That's my attitude, so!! That's what I think, I'm not scared, I don't see myself as different. To me I'm just a regular person, who's just like everybody else.
(Young male.)

For some participants, the desire to do as much as you can whilst it is still possible was not stated in a matter of fact manner as in the examples above. There was a greater sense of determination and a desire to fight the impairment:

I still go out. I'm quite a social person... ...I won't just sit there and think that I can't do this, I can't do that. To me it has to be done. I will go out and do it. It's harder. You have such fun doing the things that you want to do, why wouldn't you want to carry on doing them?
That's the way I see it. (Richard, 48, type 2.)

I went to Greece 5 years ago and I went on a quad bike by myself. They said oooh you're not allowed to do it because you've got Ushers. And I thought no, I'm going to give it a whirl, and I did give it a try and I absolutely enjoyed it. Ushers doesn't stop me from getting on a quad bike! **(I)** (Abigail, 29, type 1.)

I went on holiday to Fuerteventura, 4 year ago, with my best friend, and you know those quad bikes, I went riding with her. I wanted to drive. I didn't want Usher to stop me from doing anything. I rode it no problem. For three hours in the mountains. Volcanoes. No problem. It made feel like I don't have Ushers. We went to Florida last year. Jet ski – I rode that no problem with Abigail on the back. I like to try things. I don't want Ushers to stop us. (Erin, 30, type 2.)

These participants shared a belief in living your life and making the best from the situation, even though they knew that their vision was likely to deteriorate. However due to the lack of predictability surrounding Usher, participants are not able to 'plan for' this eventuality – if it indeed comes. Here we see that hearing and vision loss has been externalised beyond the individual, and Usher is seen as a thing at the periphery of the self rather than internalised as a way of being.

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12.2 Things that Usher stops me from doing

Regardless of individuals' determination not to let Usher stop them from taking part in particular leisure activities, Usher did impact upon people's leisure activities to a certain degree, as Sally (42, type 2) and Dave (46, type 3) discussed:

Sally: Your social life completely changes.

Dave: Aye you have to depend on someone else.

Sally: That makes it difficult, or if you are invited to parties, and if you don't know them close, I've seen myself say oh no I'm busy, because it's more hassle than it's worth.

Dave: You step back a wee bit.

Sally: I can see how you can get that you don't want to go out at all. You've got to push yourself... I love a dance, but pitch black, you can't. Your social life it changes 100%.

Likewise Bethany spoke with a degree of sadness about the sports that she had to give up playing because of her Usher:

I used to play hockey every Sunday, I played football, cricket I was captain for the girl cricket team, we played netball. Yeah hockey mainly was my favourite. I had to give all that up because it was like team sport, and couldn't really see it all to take part of it really. Yeah that was upsetting. (Bethany, 17, type 2.)

Jayne too spoke of the things that Usher prevented her from doing:

I used to go cycling but I had to give up because it was too difficult to balance not being able to see things. My balance is not very good so I wasn't able to do that. There's other things that I wouldn't want to try because I would be embarrassed because of my balance, and that stops me trying things, because I wouldn't really want other people to know that I had got Ushers. **(I)** (Jayne, 28, type 1.)

As mentioned in chapter seven on mobility, often the effort and energy used by people with Usher into getting on with daily life, particularly those whose vision loss is more advanced, also means that they do not always have the energy to be involved in additional leisure activities:

...she normally gets in half four, quarter to five, she leaves the house at quarter to eight in the morning, comes home, does her homework, has her dinner, she watches her soaps and then she's ready for bed. (Mother of Kat, 17, type 1.)

We tend to have a day off in between things. Like yesterday we had a lazy day, because we knew that we would be seeing [researcher] today. And it's the same with the comm guides, we tend to do day rest, day with them. Otherwise it's just too much. (Mother of Jess, 15, type 3)

I can do a degree of knitting and crocheting. I am happy I enjoy doing that. But there are certain things where I get tired so I have to stop. **(I)** (Susan, 48, type 1.)

Changes in the weather and lighting conditions may also mean that individuals can partake in activities one day and not another day as Jess' mother explained:

When Jess says no I can't do that my head's funny today you just say ok then maybe later, and go and do something else. And when her head's better and she stops feeling dizzy she might go on it. And they don't do that. They go come on, come on. Everybody on. Everybody. It's everybody on or nothing. So you find that a bit difficult don't you? So the best thing for you is to stay at home isn't it. (Mother of Jess, 15, type 3)

Likewise some of the other eye conditions that are related to Usher syndrome, for example cataracts, detached retinas and cystoid macular oedema (CMO) means that individuals are not able to take part in leisure activities as they so wish:

I find, because I just get so tired, just concentrating all the time, I listen to a lot of music just to relax. At the moment I'm on these Diamox tablets which is supposed to be helping out with the macula oedema. The problem that I've got. And they're taking it out of me at the moment. I'm just tired all the time. My appetite's gone a bit funny. I'm not really sleeping properly, so because of that I'm just literally resting in my room most of the time. I'll watch films. If not I'll be out of the house, because I like to be out as much as I can. I walk (guide

dog). Take her to (local park). I don't do a lot really! I wish that I did a lot more. (Bethany, 17, type 2.)

People who have cochlear implants may also choose not to take part in certain activities as Hasan's mother discussed:

I'm not sure if it will be good idea for him to be continuing competitive football, he can play for fun in teams and that, but not competitively, because you know the risk of injury, because he's got 2 implants, if he took a really bad knock on the head, I'd be really worried. And his vision, of course he realises his peripheral vision isn't that great, so umm, you know chances of him suffering an injury or tripping over the ball, I think it's something that we're going to have to see about.

(Mother of Hasan, 15, type 1.)

Faith also found it difficult to find accurate information on whether it was safe for her to do a skydive with an implant:

I'm doing a sky dive in May. It's a charity fundraiser. Before that I was wondering can I go, can I go? You can't. Why can't I? Because you've got the cochlear implant. What?! Lots of people have got implants. But the impact might damage it or something. I had to contact the hospital and find out. Can I do the sky dive? It's risky. But it's up to you. Ah what the heck! I texted people and went on Facebook, please help, and I got so many replies. I've done it, I was fine, this that and the other. And others that were ooh maybe the hospital is right. And my mum, you know mum's are always right... I know she's worried but I tried to ignore what she was saying. My friends have done it before and they've been fine. The doctor, not the hospital signed the form and was ok with it. He looked at the form, looked at the computer, and handed it to me and that was it. He didn't ask me any questions about my implant or anything. So I thought right that's fine, that's me. I'm going! I like doing mad things! **(I)**
(Faith, 23, type 1.)

However it must also be remembered that Usher is not the only thing that prevents individuals from taking part in particular activities – Ben (30, type 1) mentioned the need to save money, night clubs no longer appealed to Alex (29, type 3) and Steven (36, type 2), lack of fitness was preventing Richard (48, type 2) from taking part in cricket, Hamid (44, type 2) was involved in caring for his mother who has dementia, and health problems were preventing Brian (40, type 2), Leah (19, type 2), Olivia (54, type 1), and Victor (56, type 3) taking part in leisure activities. However the exchange between Ryan (17, type 2) and his mother recorded below raises the interesting point that people with Usher may be using other 'excuses' as to why they no longer participate in particular leisure activities:

Researcher: Are there any activities that you have had to stop doing?

Ryan: Football. Because I had my knee operation.

Mother: Plus you can't see the whole field anymore.

Ryan: I **can** just not as much... And plus I usually play about 3 or 4 hours sport. Just have one little break. Now I do about 2. Cos my leg goes all stiff, and not hurts, but goes all stiff and annoying. So I can't really bend it. And usually if I play too much sport it clicks out of place.

Similarly for some of the older participants who had significant vision and hearing loss, their preferred leisure activities were often based within the home e.g. cooking, knitting, browsing the internet. Although participants suggested that these activities were enjoyable, it raises the question of whether individuals are forced into particular types of activities, which they would not ordinarily take part in (or to such an extent) because of their Usher. Likewise some of the young people and their parents spoke about not taking part in 'typical teenage' activities due to their Usher. Leah's (19, type 2) mother said to her, "You don't go out, out. I mean when I was 16 I was out, round (city), coming home with milkman. I mean we took her (large nearby city) one night and she hated it. I mean these are 16, 18, out at nightclubs, she doesn't, she never has done." Bethany also ruefully commented that her sister has greater independence at 12 years old, than she does at 17. Callum's (14, type 1) and Daniel's (15, type 2) mothers also mentioned that their sons spent more time indoors than other lads their age.

As discussed in chapter seven on mobility, participants spoke about the lack of spontaneity in taking part in leisure activities due to difficulties with transport and getting to places. For example Usher did not stop Vicky (44, type 2) singing in a choir but it did make it difficult for her to attend practice sessions. Julie (38, type 2) enjoys swimming however she cannot get there in the evenings without a lift from her sister. Olivia (54, type 1), Alice (41, type 2) and Vicky wished that they could have more flexibility and be able to pop over and visit a friend whenever they wanted to. Participants also felt their leisure activities were less enjoyable as they used to be as they did not like relying on friends to guide them, as Nicola explained:

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Another thing that it stops me doing is going out, with mates, going to the pub, going to a club, things like that. The last time I went out was last year on my mate's hen do, and the head hen was my best mate, and she ended up looking after me the whole night. And I felt awful. She should be out enjoying herself. Watching ya. Guiding me. So yeah that's hard as well. (Nicola, 23, type 2.)

Megan also spoke of the difficulties of having to rely on her boyfriend or friends to support her whilst on a night out:

That's the way of life really. It doesn't have to be my boyfriend, it can be a friend. But if I'm out, especially if it's a place that I don't know, I **need** someone else there. It's frustrating at times – especially if you fall out with someone that's helping you! It's like I need you but I don't want to talk to you! I'd rather go out and do that, than not go out. (Megan, 24, type 2.)

Whilst recognising these difficulties a number of participants suggested that the best thing to do was to ask for and accept help or support when necessary. Michael (34, type 2) commented, "I always ask for help now cos before I didn't want any but now I just say look, if you help me I'll go, if you won't help me I won't go." However as will be discussed in the next section on adjustments and adaptations it is not always a simple straightforward task to request support. For example, working up to being able to ask for support may take time, Michael for instance said that it took him 10 years to be able to ask for support to attend the cinema.

Communication, in addition to mobility was one of the main barriers to participation in leisure activities. Rob explains his frustrations of not being able to access the information that he needed to in order to learn about growing vegetables, despite going down a number of routes to get this information:

There was one book that I really needed. It was the winter project, cos obviously you can't grow veggies in winter. I wanted to grow mushrooms. No books on audio. It was really frustrating that because it's something that I like doing, something I **really** enjoy doing, but I just can't get the books and I can't get the knowledge. The internet isn't always 100% reliable. You need expert advice which is in books. Audio books, you just can't seem to find the books you want these days. Which is really upsetting. It's very frustrating actually because I applied... there was a book and the library said to me that if you apply we might be able to get some money and get it in done into an audiobook , cos you're blind, you've got more of a need to get it in audio. OK then... sent off for 3 books. These mushroom books so I could grow mushrooms, and I haven't heard back. And that's 6 months! And I still haven't heard back and I haven't got the books. I'm really frustrated about that. You know when you want to do something and you can't get the information to do it, and nobody'll help you do it. Cos I love mushrooms! I love growing things but... the audio books are terrible, I can't seem to get the information. They really are. You just can't seem to get the books that you want. (Rob, 22, type 2.)

Olivia was also frustrated that issues regarding communication prevented her from taking part in certain leisure activities:

Also going to evening classes like a hobby, they don't provide interpreters so I feel that I'm blocked, and I can't get there. Where can you find the money for that? **(I)** (Olivia, 54, type 1.)

There are press reports about James Clarke, who has Usher, trying to claim under 'reasonable adjustments' for a sign language interpreter- but he was refused (Baynes 2012) even though he could not see to lip read or read print quickly enough.

Furthermore the nature of certain leisure activities makes communication more difficult. Participants spoke about pubs and cinemas being dark which makes lip reading and sign language more difficult. Rosie (21, type 2) who usually communicates using spoken English explained that she used the deafblind manual alphabet whilst at

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the cinema as it is too dark to lip read. Particular sports or rides at theme parks may mean that hearing aids, cochlear implants, or glasses may not be able to be worn. Ryan for example complained that he was not able to play rugby because of these reasons:

Rugby. Cos if I take my glasses off I can't see. And I have to take my hearing aids off because I don't want to ruin them. Cos I love rugby so much. It'd just be hard. (Ryan, 17, type 2.)

Swimming in particular was mentioned as difficult. Brian (40, type 2) had given up swimming after an incident when he was made to feel embarrassed because he could not hear what the lifeguard was saying. Callum's (14, type 1) mother said that although her son loved swimming he had to give up because he just could not communicate with his hearing friends once he removed his cochlear implant. This left Callum feeling vulnerable, especially whilst in the water. Jess (15, type 3) had also given up swimming because she found it too scary and difficult because of her hearing and vision loss, however over time Jess had begun to get back in the water and had started attending training sessions. Jess usually used spoken English to communicate however having to remove her hearing aids meant that she used hands on signing whilst at the pool. As well as having to deal with this change in communication method it meant that her mother always had to support her as at the present moment Jess' communicator guides were not able to use hands on signing.

As we shall see in the section on leisure activities specifically for people who were deaf, visually impaired, or have Usher, communication and mobility issues are two of the reasons why participants in the project suggested that at times they liked to join in with people who are similar to them. However we shall first look at the adjustments and adaptations adopted by people with Usher in relation to leisure activities.

12.3 Adjustments and adaptations

As seen in chapter one on diagnosis, adjustments and adaptations are a feature of the lives of people with Usher. This is due to the progressive nature of Usher syndrome. When we speak of adjustments and adaptations they refer to both the psychological and practical

adjustments and adaptations that people make. Linda (46, type 1) for example had to psychologically adjust to the fact that she could no longer ride a bike, as well as making the practical adjustments in finding information out about her local tandem cycling club. One way that participants adjusted to not being able to take part in leisure activities was to focus on what was possible rather than rueing what was no longer possible. For example Victor (56, type 3) commented, "There are certain things that I just can't do anymore. I try to concentrate on the things that I can do." Likewise Alice said:

I used to love, maybe on a Friday evening, going to meet some of my friends, go for a pint, which I don't do anymore, cos of the lighting. Shopping, I used to love. Clothes shopping now, you go into a shop, and the lighting it's very dull, so I don't enjoy shopping anymore. I tend to do my shopping online now. Cinema I used to love, but not anymore, cos corners are cut out. But just get on with it, that's my motto. (Alice, 41, type 2.)

In order to make adjustments and to adapt to new ways of doing things participants need awareness of different approaches. For example a number of participants spoke about how the Paralympics had inspired them to start, or start to think about, taking up sports, as Vicky explained:

And actually what me and my husband have started doing, actually that's not us alone, we've started going running, but holding hands now in the dark. Now I know it sounds a bit soppy and romantic, but I got that idea from the Paralympics. (Vicky, 44, type 2.)

Similarly Ryan (17, type 2) due to his awareness of goalball suggested that he would continue to play football in the future, "Cos if my eyes do get worse I'll still play football. So... I'd play blind ball instead... So that's alright. It's not that much difference." However he was unsure about playing tennis and competing in athletics as he did not think that he could 'because of his eyes.'

Participants tended to be more accepting of no longer being able to take part in certain leisure activities if they saw Usher as a problem with their hearing and/or vision which, if fixed, would allow access. This individualistic understanding of Usher was presented by Ben:

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I would have liked to have done karate, I know that I would have been good at it. I would have been committed. I would have loved to have got a black belt, and trophies, but I just had to give it up. Because obviously somebody could hit me in the face, and I wouldn't be able to defend myself. **(I)** (Ben, 30, type 1.)

Likewise Jill (59, type 1) in one of the focus groups, listing characteristics of herself suggested, "I'm a friend. I'm excitable. Self-motivated. I'm optimistic. If it wasn't for my eyes I would be a traveller." However participants who saw the barriers to inclusion as external to themselves, for example related to mobility or communication, tended to be more focused on making adjustments and adaptations and doing things differently. Jess (15, type 3) explains, "I just need to do it in a different way to what someone else would. I can do whatever I want, just not always the same way as someone normal." Kat (17, type 1) for example was no longer able to keep up with dance classes with her peers so instead enjoyed dancing at home with her sisters. With regards to Ryan's sporting achievements and his dedication to carrying on as much as possible albeit differently, his mother comments:

But they all think you're wonderful because you still do it, don't they? They admire you because he can't do something, but he still gets up and has a go, and if he can't do it he can't do it. He just finds a different way of doing it. He's clever. Oh don't sit there and start bawling! I'll have to go and get a tissue! (Mother of Ryan, 17, type 2.)

As will be discussed in the next section, one of the main ways that people decided to do things differently was by joining a group specifically for people who are deaf, blind or have Usher. Another common way to adapt to a situation was to enlist the support of other people – going to the gym with a friend and making sure that you were not left alone whilst at the pub were commonly given as examples. As mentioned above, it is not always easy to ask for support, and may take an extended period of time to pluck up the courage to ask for support, nevertheless this was seen as a simple solution to carrying on with life. It is worth noting that it may be easier to ask for assistance with regards to leisure activities than for other activities, such as household shopping or child care, as leisure activities are often frequently undertaken with other people, and often a closeness of bodies more acceptable.

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The majority of participants however did not solely inhabit one side of the divide between individualistic and externalised understandings of Usher. Chris (33, type 3) for example spoke of not taking part in regular Sunday league football with his friends and neighbours as his visual impairment would make it too difficult to join in, however with regards to cricket he has made adjustments and now regularly plays blind cricket at an international level. Susan (48, type 1) explained that she could no longer go shopping independently due to being deafblind, however organising her kitchen in a specific way, and labelling cupboards and ingredients with braille meant that she could continue her hobby of cooking. Such approaches demonstrate the multi layered approaches of living with Usher that the participants in the research project adopted.

12.4 d/Deaf, blind and Usher specific leisure activities

As suggested above and also in chapter nine on family, friends and social networks, some participants in the study enjoyed meeting up with other people who were d/Deaf, had a visual impairment, or had Usher.

One of the reasons that participants enjoyed contact with other people like them in relation to their hearing and visual impairment was that it made communication and mobility easier, which as suggested above were two of the biggest barriers to participation in leisure activities.

The desire to communicate with peers is demonstrated by Faith's willingness to travel many miles to catch up with other BSL users of her own age:

...there's so few youngsters in the area. But sometimes I'll travel to [city 150 miles away] and we'll meet up and have a drink and a blather. You know because [home town] has a lot of older people, and I'm not one to socialise with the over 60s unfortunately **(I)**. (Faith, 23, type 1.)

The experiences of Olivia (54, type 1) and her sister Gill (59, type 1) and their friend Barbara (50, type 1) highlight that due to the relatively small number of BSL users in the UK and the fact that around 5% of profoundly deaf people are likely to have Usher (Millan et al, 2011) it is

not always possible to distinguish between Deaf people getting together, and people with Usher getting together:

Researcher: Do you meet because you are friends or because you have Usher?

[Barbara: That's a good point. I think we're friends. It's nothing to do whether we have Ushers or not. (I)]

Olivia: yes and my husband is friends with Barbara's husband. And we meet up. Go out. We were friends before the Usher thing came into being. (I)]

Mothers of the young people in the project spoke of being in contact with the National Deaf Children's Society (NDCS) and joining in with social activities such as trips to theme parks, picnics and holidays. Some of the young adults also remembered taking part in NDCS activities when they were younger. Although centring round 'fun activities' these events are focused on information, support and knowledge sharing as Ryan's mother explains:

He was quite paranoid. Then we joined that Deaf Children's Society and he went to (theme park) and he was actually amazed at how many children wore hearing aids. And it made him feel more comfortable. (Mother of Ryan, 17, type 2.)

In general however, participants who were not BSL or SSE users did not mention a desire to socialise with people who had a hearing impairment. This may be due to the fact that it is a shared language that brings Deaf people together, regardless of hearing status . Consequently spoken English users share a common language with non-hearing impaired people, so there was not a desire to join together in leisure activities with other hearing impaired people. Chris explains his location within visual impairment rather than hearing impairment:

I don't need to be in the Deaf world for communication purposes, because I'm coping well enough as I am now, but I need to be in the visually impaired world because I love sport and I want to play sport at an international level... but it could have been different... I might have ended up in the Deaf world, just because that drive would have

been about me accessing communication, rather than accessing say, playing sports. (Chris, 33, type 3.)

In the same way that groups for Deaf people tended to attract people who face the biggest communication challenges, groups for the visually impaired tended to attract those who face the biggest challenges with regards to mobility and taking part in their chosen activity. Michael commented:

It's more harder hanging out with people who have no visual impairment, because they don't really understand. They tend to leave you behind. They don't mean to, it's their natural way of doing things. (Michael, 34, type 2.)

Groups which join together around visual impairment provide the appropriate adjustments and adaptations needed in terms of equipment, specialist advice and coaching. Rob for example was excited about learning archery:

I don't know what to expect. I don't know how it's done. I know that they line you or something then you fire. That doesn't help when you can't see the target! And I'm more worried about hitting another blind person – you know what I mean. (Rob, 22, type 2.)

Activities specifically for people with visual impairments allowed, among others, Michael, Alex, and Chris to play cricket, Victor to go rowing, and Rosie to play goalball. It is worth noting that the leisure activities for people with visual impairment that the participants took part in centred around sports, no one spoke about arts groups for people with visual impairments. However as would be expected for people with Usher the combined nature of hearing and visual impairments cannot be ignored:

...about ten, eleven years ago, I went with a blind group bowling, there was a communication breakdown and people didn't seem to understand about Ushers so I sort of stopped that because of the communication breakdown. (Brian, 40, type 2.)

Likewise Kat (17, type 1) was having difficulties at her climbing club for visually impaired people as she could not hear the directions from her coaches.

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Nevertheless no one spoke of joining in leisure activities specifically for people with Usher. Although socialising may be a by-product of these activities, Usher events, like NDCS events, tended to concentrate around information and support, much to Michaels' chagrin:

I did feel more comfortable with the Usher group. We did meet up. Went to London and some posh hotel thing there. 17 quid for two glasses of wine!!... All they do is sit down and talk about Usher, I didn't really want to, I know I've got Usher but I just want to come and meet people and have a laugh. It's good to talk about it the first time, but next time it's more, like speeches (makes snoring noises) – I've had enough! I'm depressed enough – I don't need to talk about it! I just want to go out. They said that there was all these activities, and they probably did, but I never knew about them so... I want to go to groups but I don't want to talk about it all of the time. I know I've got Ushers! (Michael, 34, type 2.)

As was shown in chapter nine on family, friends and support networks the importance of Usher networks are not to be underestimated in relation to support and information; however with regards to leisure activities it appears that Usher specific groups are less important. The most important thing was that participants were able to get on and do the activities that they wanted to; life does not always need to be focused around Usher.

12.5 Discussion

12.5.1 Change

As with other areas, Usher brought change to the leisure and free time activities of the people in the project. They had to adapt to changes whether they wanted to or not; and whether they realised that the changes were related to Usher or not. Change in leisure activity is of course something which happens to most people as they grow, and then grow older. Interests change and abilities develop, or perhaps reach a peak and then fade away.

Some people spoke particularly about changes to leisure which had happened specifically because of the activity and Usher – such as

involvement in sports (Bethany was sad that she had given up the competitive games she had previously been involved in because she could not see). In other cases, change was not because they could no longer participate in the activity, but because other aspects of Usher affected it – the change in communication needs, the change in mobility, which meant that they could not get to activities or communicate with people (for instance, Vicky wanting to visit a friend, Brian going to bowling). Using the swimming pool raised some difficulties because people were able to swim, but could not manage the communication around swimming when they were no longer able to communicate without their hearing aids or implants. There was also a significant change in levels of fatigue brought on by the need to concentrate and be alert, (even within social and leisure activities) at a much higher level for people who have to deal with reduced vision and reduced hearing. Bethany explained how she had to concentrate all the time, and so was tired. Susan mentioned getting tired when she used her eyes.

Participants did mention some changes that other people had made to adapt for them, but there was also mention of situations which could have been improved through changes in other people. For instance, Sally talking about going to parties says that she does not go, because 'it's more hassle than it's worth'. The social environment in which someone automatically thinks about and then compensates for this difficulty (for instance by offering transport in both directions and thinking about what will happen at the party that Sally could join in) is perhaps a forlorn hope. Improvements in communication and understanding in other groups though could have helped Brian to continue going bowling (he stopped because of communication difficulties). Sharing information about what could happen had helped some – for instance, Vicky and her running hand in hand, once she had the idea. Ryan might have found information about goalball helpful, and better information about tandem clubs would have helped Linda.

12.5.2 Predictability/uncertainty

A number of participants continued with activities or even started them because they did not know when they would lose vision and so stop being able to. This meant that people did things which they might have left till later or got on with it and did things now.

Leisure Activities

They were also uncertain what they could, and could not do, and what they might be allowed to do or not. Uncertainty was not only from the participants themselves, but also doubt about whether they should do things (such as Faith skydiving and Abigail on a quad bike). Richard was not sure if he'd manage a 4 hour walk, but then was pleased when he did. In other cases, there was uncertainty about what might be available to them, - Rob's experience with libraries was very frustrating for him, when he wanted a talking book about growing mushrooms.

The uncertainty about environments was shown by participants who would have liked to go to the cinema but were unsure that the environment would allow it – darkness meant they could not move about or communicate effectively. Pubs and clubs are also dark and there were similar considerations.. People with disabilities frequently have to face unpredictable environments, but for those with Usher, as they lose their vision, they notice that their hearing does not function as other people's does – which has not been an issue for them before.

12.5.3 Diversity

Despite all this, there was a wide range of leisure and social activities being undertaken by the participants and with a wide variety of others. Sports activities were common, but there did not seem to be many limits on what people would attempt, though some activities needed to be adapted for visually impaired people (such as running holding hands or playing blind cricket). The difficulties with swimming were not related to swimming itself, but to the surrounding aspects of communication and information. While vision appeared to be the problem for other activities, it was not always visual loss that was the most significant factor – communication was what stopped Brian bowling.

Some people were ready to travel distances to events (sporting and otherwise) to meet other people, including those they found they had something in common with – Faith travelled 150 miles to meet other BSL users. There were adventurous spirits quad biking (Abigail) and skydiving (Faith) , going on the roller coaster rides, tandem riding (Jess) and more. Others preferred quieter activities, such as knitting, (Olivia), gardening (Brian and Rob) or music and TV (Bethany). While we did not ask for specific measurements of visual and hearing impairment, it did

appear that those with higher levels of vision and hearing loss were more likely to be involved in activities closer to home.

Some people happily asked for help from friends – Michael describes how he built this help into his arrangements. Others however did not want to be dependent on their friends or to use their peers as guides (Nicola talks about this).

There was a range of things which people had stopped doing, or did not take up and some activities other people *did* take up – such as tandem riding, instead of a solo bike, or going to the cinema with a friend instead of alone. Some still liked going shopping, while Alice found it difficult, and now shopped online (compare to Rebecca Alexander who moved to New York where *everyone* of her generation shops on line – so she does not feel different). While Ben decided not to take up karate, other participants were involved in a range of sports, including martial arts which involved more contact. For some Usher might be a reason to stop doing things, while for others it might be more of a challenge – as Ryan’s mum said “he still gets up and has a go, and if he can’t do it he can’t do it. He just finds a different way of doing it”

13 Self Image Profile (SIP)

In addition to answering the interview questions, participants who took part in the initial interviews were asked to complete a self image profile (SIP). The results from this survey, will be presented and discussed in this chapter. However, before this we shall explore what SIP is, why the self image profile was beneficial to the research project, and finally how the SIP was administered.

13.1 What is SIP?

SIP is a standardised assessment tool used to measure self reported levels of self image and self esteem. The self image profiles for adults were created by Richard Butler and Sarah Gasson with East Primary Care Trust (Butler and Gasson, 2004). The self image profiles for the young people were also developed by Butler and Leeds NHS Trust (Butler, 2001). Self image “reflects how *I think about myself*, i.e., how an individual describes or construes him/herself”, whereas self esteem “implies an evaluative or affective judgement and reflects how *I feel about myself*” (Butler and Gasson, 2004: 3). Self esteem is the perceived distance between how an individual sees themselves and where they wish to be (Butler and Green, 1998). The SIP for the young people measured positive and negative self image (Butler, 2001). A positive self concept is linked to higher levels of well-being, academic achievement, autonomy (Butler and Gasson, 2005).

The SIP is a standardised tool within a British context, and we can compare scores across the general population. However we could not find any evidence of it having been used before with d/Deaf people.

There are problems associated with SIP as it relies on self reporting and presumes that we can quantify self image and self esteem (Butler and Gasson, 2005.) Also since the survey was administered after the interview, perceptions of self may have changed as a result of the interview. Nevertheless the SIP provides a quantifiable snapshot of how participants thought about themselves. For the adults, it also showed how far they felt from their ideals.

13.2 Why SIP?

We chose to use the SIP, rather than any other standardised measures of self image and esteem that are available, because the latter tend to focus on the negative aspects of people's lives – of what they can and cannot do. We did not want to hear only about the difficult things; we wanted to know the good and positive things as well. While the SIP is a standardised measure, our use of it was not be standardised. It was delivered in other formats, including in BSL (see below). A very significant reason for choosing it was that it could be presented in BSL without a great deal of translation and that it was not over wordy; we believed it would be suitable for those whose first language is not English or for those who may be at the early stages of reading braille.

However, as it is a standardised measure on a British population, it allowed us to compare our results with a wider population. Studies such as Bodsworth et al (2011) and Wahlqvist et al (2013) indicated that our participants could show considerable difficulties with how they thought about themselves. This measure allowed us to have more than just a narrative perspective on the lives and experiences of individuals.

Our sample of 41 participants gives some interesting insights into how people see themselves. It shows a picture of individuals who have different thoughts and feelings about themselves and who have different feelings about their future –sometimes positive, sometimes not.

13.2.1 Administration of the SIP

41 of the 42 participants who took part in the initial interviews completed the SIP. We used both SIP scales, the one for adults and the one for young people. Consequently we have responses from 30 adults and 11 young people. Participants were asked to rank themselves, on a scale of 0-6, on a list of different characteristics, where 0 is 'not at all like me' and 6 is 'very much like me'. The different characteristics included things like happy, friendly, hard working, and confident. For adults, all the characteristics are positive while for children, about half are negative – things that it would be expected they did not want to be – bossy, worry a lot, shy. Participants had to do the survey twice – the first time they had to say 'how you think you are', the second time they had to say 'how you

would like to be'. The second time the scores could be higher, lower, or stay the same. It was up to the people taking part to decide if 'how you would like to be' meant with Usher or without. This is because some people cannot picture life without having Usher, or because Usher is such an important part of who they are that if they excluded that, they would be excluding an important part of their identity.

The outcome measures for the SIP are a self image score and a self esteem score. The self image score is calculated (for adults) by a total of their scores of 0-6 in relation to the 30 items. It assumes that 6 is a score indicating high self image. The self esteem score measures the difference between the self image score and the score (on each item) for how they would like to be. A good self image is where the two scores are similar – they are how they would like to be.

For adolescents, the test includes 12 positive characteristics, 1 neutral (whether they feel different from others) and 12 negative characteristics. The two areas – positive and negative – are scored separately, giving two scores for self image (positive and negative) and then two for self esteem.

Participants accessed the SIP in the most appropriate way for them, including standard print, large print, coloured paper, using a handheld electronic low vision aid, by the researcher reading out the questions and filling in each participant's answers, and on the computer with screen-reading software. Braille copies were available but participants preferred to use other means. The majority of participants enjoyed filling in the SIP, and those people who had been a bit shy, or felt that they did not have that much to say during the interview, still filled in the survey, and often got a sense of satisfaction in completing it.

13.2.2 Adult SIP

30 adults completed the SIP – 10 were men and 20 were women. 16 were in the younger adult age group (19 – 35 years old) and 14 were in the older adult age group (36 – 56 years old). There were responses from 9 people with type 1 Usher, 17 with type 2 Usher, and 4 with type 3 Usher.

Overall, when comparing participants' responses with the general population the vast majority of participants' scores were within the normal range.

The SIP scoring gives different cut-off points in relation to the normal range in reference to people's age and gender. Consequently this discussion will also look at gender, and age, as well as looking at people with different types of Usher.

Adult participants ranked themselves on 30 individual characteristics. Overall the 5 characteristics that participants were most likely to see in themselves (in descending order) were honest, trustworthy, friendly, loyal, and caring. Participants saw themselves as least likely to be (in descending order) thin/slim, optimistic, fit, patient, and confident.

By comparing the scores for what they saw in themselves and what they would like to have scored, we are able to discuss how content they were with their own profile.

Participants were more satisfied (in descending order) about how happy, honest, trustworthy, friendly, and loyal they were. Participants were less satisfied (in descending order) with how sensitive, thin/slim, generous, good listener, and patient they were. The gap between how people see themselves and how they would like it to be was smallest (in descending order) in relation to these characteristics – good listener, honest, caring, friendly, kind, and loyal. The gap between how participants are and how they would like to be is largest (in descending order) in relation to these characteristics – sensitive, thin/slim, optimistic, fit, confident.

Examining these characteristics, 'sensitive' is particularly interesting. The characteristics are presented in such a way that participants generally indicated that they wanted to become more like a particular characteristic – they would like to be happier, more optimistic or more honest than they rated themselves (this is the way the profile is supposed to work). For 14 of the 30 characteristics presented, all 30 participants wanted to be at a higher score level for each of these. For a further 12 characteristics all participants bar one, wanted to become more like this characteristic. For three characteristics, only two or three people wanted to become less like this characteristic. However nine of the 30 participants wanted to become **less** sensitive. Of these nine

Self Image Profile

participants, eight were women. They were mixed in terms of Usher type and age. This finding may be related to a discrepancy in understanding what is meant by sensitive – for example it may mean someone who thinks about other people, someone who is overly responsive to other's feelings, someone who is allergic to things, someone who gets upset easily. Furthermore sensitive can often be seen as a negative characteristic, especially when applied to females.

If we return to the interviews we see that Vicky (44, type 2) who in the SIP said that she wanted to become less sensitive, also spoke of wanting to become less 'emotional' with regard to her Usher, and was taking part in mindfulness courses in order to discover ways of channelling her energy in positive ways. Victor (56, type 3) the only male to want to become less sensitive also took part in mindfulness courses. Similarly Sally (42, type 2) who also wanted to be less sensitive, spoke in her interview about her worries of being seen by other people, because of the 'hidden nature' of Usher, as a fraud, but at the same time suggested that a person with Usher just has to ignore these people. It is interesting to note however that it was males who were more likely to display emotion e.g. welling up, voice cracking, changing of the subject, than female participants.

In looking at the scores of siblings who completed the surveys we see that they often had different scores, highlighting that people do have different responses to Usher even if they generally share the same background and genetics.

The 30 characteristics were grouped into six categories in order to assist with analysis of aspects of the self. The categories were ; Outlook, Consideration, Social, Physical, Competence and Moral. The characteristics included are listed in the table below (patient and friendly were scored twice; enthusiastic was not scored.)

Outlook	Consideration	Social	Physical	Competence	Moral
Happy	Patient	Friendly	Fit	Creative	Hard working
Optimistic	Caring	Sociable	Active	Organised	Loyal
Easy going	Good listener	Fun	Thin/slim	Determined	Trustworthy
Patient	Thoughtful	Outgoing		Intelligent	Reliable
	Helpful	Sense of humour		Confident	Honest
	Generous				
	Sensitive				
	Kind				
	Friendly				

Participants were asked to rate themselves as on the scale from 0 (not at all like me) to 6 (very much like me). On average, based on these ratings, participants were mostly likely to see themselves as most like the characteristics in the moral category (4.9) followed by consideration (4.6), social (4.3), competence (4.1), outlook (4), and finally physical state (3.4). (The higher the number, the more like that characteristic they were). If we look at these characteristics based upon Usher type we see that scores are pretty consistent with the overall scores for the categories of consideration, social and moral. However if we examine the Outlook category we see that on average people with type 1 Usher scored 3.6, people with type 3 Usher 4, and people with type 2 Usher 4.2. Although the numbers are small, this does suggest that people with type 2 Usher may be happier, more optimistic, easy going and patient than people with type 1 Usher, or at least report themselves as such. Why this might be is a question which this study and this profile does not answer. Again, the numbers are small, but on average, younger adults had a higher score on the 'Outlook' questions – with younger adults scoring 4.2 and older adults 3.8. However in relation to the types of Usher this cannot explain the answers as the split of younger and older participants is roughly equal in all 3 groups.

The category Physical also shows a difference in scores, but a different one. People with type 2 Usher scored 3.1 in this category, people with type 1 Usher 3.7, and people with type 3 Usher scored 4.1. Thus those with Usher 3 considered themselves the most physically fit, and those

Self Image Profile

with Usher 2 the least. It is not because all the people with type 3 Usher were men, because if we look at the scores, men and women are similar at 3.5, and 3.4, respectively. This is interesting in the light of the fact that no one, regardless of their Usher type said that they were unable to take part in sporting activities because of their Usher. It is interesting also in the light of the balance problems usually associated with Usher 1.

Competence is another category where we see differences in the scores of people according to their Usher type. People with type 2 Usher scored themselves at 3.9, people with type 3 Usher scored 4, and people with type 1 Usher 4.4. Again men were less likely to see themselves as competent, scoring 3.8, as opposed to the women's score of 4.2. This may go some way to explaining the higher score for people with type 1 Usher as this contained only one man. However, as mentioned previously all the people with type 3 Usher were males.

If we look overall at the three adults who scored the highest and lowest in terms of self image and the highest and lowest in terms of self esteem we see no patterns according to age, gender or Usher type, except in relation to self esteem and age. All the participants who ranked the three highest scores in relation to self esteem were in the younger adult age group, and the three who scored the lowest in terms of self esteem were all in the older adult age group. The two lowest scores for self image included two of the lowest three in terms of scoring in self esteem. Since we did not use any objective measures for vision and hearing loss, it is not possible to compare such measures to the self image profile, but it appears that those who had very low vision, or profound hearing loss were not less likely to have good scores on the profile.

The three highest scorers overall did have the highest score for outlook and the three lowest scorers overall had scores for outlook in the lowest three. The three highest scorers overall also scored in the highest three for the social category, and of the three lowest two had the lowest three scores for the social category. The outliers are also interesting; although of course they could just relate to participants tiring or not understanding the questions. One person who scored in the lowest three for outlook also scored in the highest three for the social category. One person who scored in the lowest three for self image scored an average for self

Self Image Profile

esteem. One person who scored at the lowest level for physical and for moral categories scored at a high level for outlook. While two of the lowest three scores for self image also scored within the lowest three for self esteem, none of the three highest scorers for self image scored at the highest level for self esteem.

The SIP profile for adults provides cut-off scores – the figures which would indicate ‘an unusual profile’ and which might lead to further action. Of the SIP self image scores, only one person fell outside this bracket. This person also had a low self esteem, though only just outside the cut-off figure. No-one else’s self esteem figure fell outside the figures which would indicate an unusual profile – that is, reach the cut-off figures which are set at two standard deviations beyond the mean from the figure for the general population.

21 people scored at 3 or higher on the question about optimism – that is, they were midway along the scale for optimism. Of the 8 who scored 1 or 2, one of these only considered that they would like to be at 3 (moderately optimistic) whereas all the others said they would like to be very optimistic (i.e. they would like to have scored 5 or 6).

13.2.3 Self image and young people

The self image profile for young people has different categories and key words. The words in the adolescent profile include negative phrases such as ‘bossy, mess about, worry a lot’ unlike the adult version. The participant achieves a positive self image score, a negative self image score and a discrepancy score, related to how different they are from how they want to be.

Eleven young people completed the SIP for adolescents. There were responses from six females, and five males. Six people had type 1 Usher, four people had type 2 Usher, and 1 person had type 3 Usher. While a study by Van Gent et al (2012) using another measure suggests that deaf adolescents have more problems with social acceptance and with close friendship than comparable hearing peers for this measure, all the participants scored within the normal ranges expected of their age and gender. For the young people because the sample was so small it was not possible to look at differences according to gender and Usher type.

Self Image Profile

The 25 characteristics were divided into three; 12 characteristics indicative of positive feelings and 12 of negative feelings. There was then one category 'Feel different from others' which was neither of these – a measure of the 'sense of difference' or separation between themselves and others.

Positive	Neutral	Negative
Kind	Feel different to others	Lazy
Happy		Annoying
Friendly		Moody
Funny		Mess about
Helpful		Shy
Hard working		Cheeky
Talkative		Loud
Confident		Sarcastic/bitchy
Sporty		Worry a lot
Intelligent		Bossy
Fun to be with		Short-tempered
Good looking		Get bored

The characteristics that the young people saw themselves as being most like were (in descending order) friendly, helpful, fun, worry a lot, fun to be with. Participants were least likely to view themselves as (in descending order) bossy, sarcastic/bitchy, annoying, mess about, moody. The characteristics that participants most wanted to be like were (in descending order) fun to be with, friendly, funny, helpful, kind. The characteristics that the young people least wanted to be like were (in descending order) bossy, annoying, short tempered, shy, moody. The gap between how participants are and how they would like to be was smallest (in descending order) in relation to mess about, sarcastic/bitchy, friendly, helpful, loud. The gap between how participants would like to be and how they are was largest (in descending order) in relation to shy, worry a lot, get bored, confident, short tempered.

We see that the participants were most likely to see themselves in positive terms. However 9 of the 11 young people said that they worried a lot, and seven out of 11 said that they would like to worry less. The worry figure is odd, though, because some of them said they would like to worry a lot – and this, if true, would be very unusual. Five out of the 11 young people said that they did not feel confident, and seven of them

Self Image Profile

would like to feel more confident. Six out of the 11 said that they were shy, and 8 of the 11 would like to be less shy. This did not necessarily come across in the interviews, as often the young people were telling us about the fun and exciting things in their lives.

Also we do not know if their worries, or shyness, or lack of confidence was related to their Usher – it may have been because of other things such as friendships, relationships, or issues at school. Therefore it is useful to combine the SIP and interview to provide a more rounded account of people's experiences. It may also be part of being human to want to change and develop yourself, because sometimes even when the young people agreed with positive characteristics they still wanted to score higher in this category. For example all 11 young people said that they were friendly yet three young people still wanted to become even more friendly.

One of the questions asked the young people to score themselves on 'feel different to others'. It was expected that participants with Usher would feel very different from others, and would want to feel not at all different to other people; however, the results were surprising. Two young people felt very different scoring a '5' and a '6' and wanted to **remain** feeling very different. Two other participants felt moderately different and wanted to **become more** different. Only two from the 11 young people reported feeling very different and wanting to be not at all different. Also if we look at the overall scores of positive self image we see that of the two people who scored most highly in relation to self image; in relation to being different one ranked themselves as '6' and wanted to be '2' and the other was '3' and wanted to be '6'. This was not consistent with those with having positive self image feeling the least different to other people. If we look at the scores for the two participants who had the lowest scores for positive self image we see that one does follow the expected trend by scoring themselves as a '5' and wanting to be a '0' but the other person went against this, suggesting that they did not feel very different at all with a '1' and wanting to become a '0'. Likewise the two participants who had the lowest levels of negative self image wanted to become a '5' and a '6' in relation to feeling different – they wanted to be different from others. The two participants who had

the highest levels of negative self image scored themselves 'said they did not want to be different (they scored '2' and '1').

13.3 Discussion

While studies have shown that people with deafblindness, and people with Usher can be expected to have significant problems in relation to depression and pessimism, the results from the SIP with both adults and adolescents do not demonstrate such low self image as might be expected. They are happier – all of them scored themselves as between 3 and 6, with 8 at point 3 and 7 at point 6 - than might be expected for people with Usher. They mostly considered themselves to be fun (24 scored 4 or above in this category) and active (19 scored 4 or above in this category). 21 even rated themselves as moderately optimistic. Their outlook scores as a whole reflected some positives; 16 scored 4 or above on all the aspects of outlook and 10 scored between 3 and 4. Four scored less than three, of whom one scored less than two. However, it could not be said that they saw themselves as depressed.

It is important to remember though that this scale is a self report. They had just been talking about their lives and the things they were doing and this could have led to an unduly positive estimate of their typical feelings. The scale does not ask about whether they have ever been depressed, or how deep such depression might have been. The young people's scores in relation to worry are difficult to explain – it is odd to think that they would want to worry more. How this might relate to Usher is unclear. (Their scores in other areas on the test do not suggest they did not understand how to complete it).

The profile also depends on the ability of the person to think that things could change. If they did not think anything could change, then they were likely to score the items both times in the same way, which would lead to a high self esteem score (though it could still be a low self image score). For this reason, among others, the self image measure was more useful overall. There was also a concern that some people might not want to *think* about how they could be different. Since admitting that there was the potential to change themselves might mean they had to do something differently, they may have simply scored all the items as the

same for how they are now and how they would like to be. More men than women scored items more closely – although we did not measure how long it took people to fill it in, women seem to have pondered longer over the options.

Overall then, the Self image Profile was an interesting way to learn about the young people and the adults we were working with. In one sense, it was taking the temperature as an absolute measure, rather than an overall consultation. It represents how they felt about themselves at that moment and the scores cannot be causally linked to Usher, or to their other interview answers.

One person scored at lower than usual score in relation to both self image and also had a low self esteem score. This participant was signposted to other agencies that may provide support for people with sensory impairments. One person had a lower than average self esteem score. This person had requested further details from support agencies during the interview and consequently these were provided. One person also had a lower than average score in relation to self esteem. However on reflection this person's score was only one point below the 'normal' cut-off point and having seen the strong family and social networks that were available to them it was decided not to signpost this individual to additional agencies.

The test was a good choice in that it was simple to administer, did not require lengthy explanation and was not wordy. Most people seemed to enjoy filling it in and it has given us some quantitative material to add to the interview data.

14 Discussion

14.1 What is it like to have Usher in the UK today?

At the end of the study, we look back and reflect on what it is like to have Usher, today, in the United Kingdom. From the voices of people with Usher themselves we can see some things emerging

14.1.1 **They enjoy life**

From some of the literature (e.g. Miner 1995 and 1997, Bodsworth et al. 2011) we had envisaged the possibility that most people with Usher might be living with high levels of depression, feeling unconnected to society, unable to think about the future and unfulfilled and unsuccessful in life. But this was not the case. We also found excitement, interest and anticipation. People with Ushers were not unusually pessimistic (the SIP told us this). They were doing things they wanted to do, either despite, or occasionally because of, Usher. They were successful (if sometimes worried) parents, competent employees, and good friends. They played music, went to parties, and took part in sport. They found friends or supporters who also had Usher or joined Facebook groups. They had already accepted being deaf – becoming visually impaired was not something they wanted, but it was the hand they had been dealt. Usher was obviously part of what was going on, but it was not all of it. They did most of the things which other people in the UK did. Undoubtedly some were, or had been, depressed. Depression may have occurred when they heard about Usher, but it also happened at moments of change, particularly those associated with mobility – such as starting to use a cane, or giving up driving.

14.1.2 **They manage**

Like many other people in the UK, they learned to manage, and they managed. Having Usher did mean that they had to do things differently, and perhaps to give some things up. They had to learn to use specialist aids – maybe a cane, or an interpreter, or work out what a communicator guide could or could not for them. They found ways of using technology to help them (not by any means always specialist technology). They found ways to get where they wanted to go using public transport, or

using a guide dog, rather than having the independence of driving themselves. They used various types of support, when it was given, and learnt new skills, from learning a new job to practising mobility routes with a specialist. They continued to learn (if at school) and found out how to tell teachers to get it right for them.

Interestingly one of the main things they did was **turn on the lights**. Obviously it is important that such lighting did not create glare, but lighting played a very significant part in people's ability to manage.

Sometimes of course, they did not manage. There were understandable and often avoidable frustrations. Some of these were with people; with colleagues who 'forgot' they had a visual or hearing impairment, neighbours who might think they could see if they did not use a cane. Some of these were with systems; they could not get a bus pass which let them go to work, they were not provided with good information at diagnosis, their equipment was not appropriate for their needs. Sometimes the environment was against them; trailing branches, poor lighting, lots of people.

14.1.3 **Things develop**

For each individual things developed and changed but they also developed within society. Change is part of the natural process of life that things change. While it is certainly true that things may have changed a lot for some of the participants, change is not, of itself, unusual.

What was more unusual perhaps was that the changes challenged their own perceptions of themselves, that they challenged individuals' ontological security. Not knowing when things will change, and how much they will change means that they live sometimes with little **continuity** - a defining characteristic of this security. As they live with Usher, other things will change too, and those changes interacting with the difficulties of having Usher- moving house, having children, losing a job, make that continuity very hard. This unpredictability of life was reflected in people's accounts of 'doing things while I still can' and 'I don't know if it's worth it..'

Discussion

Unlike having diabetes or cancer, no new routine is set up (medication, blood tests, hospital appointments). People have to live their lives, in new conditions, and in many cases, without much support. As Julie said

Sometimes I feel like we don't get as much support compared to ... other people that have other things wrong with them. My friend had a form of skin cancer,, and it's bad, but she's got the all clear now, but when she were diagnosed she got so much support, she got offered counselling, she even got offered hypnotherapy, and things like that. I had nothing. I didn't get anything at all.... if you've got diabetes..., you get this and this, and you get a diabetes specialist nurse, who comes round to see you and all this; you know it's ...all a bit in a grey area. I think for Ushers, It just don't seem to be something that needs a lot of support. I don't know why. Maybe because it's not that common?

For people with Usher, there will be no 'all clear'. They will not get better, and no matter what they do, they will in fact get worse.

Through the cross section of time which can be seen by interviewing people of different ages, we see changes in people but also in society. Three of these which are especially visible in this study are changes in the identification of the typology of Usher, changes in education and changes in diagnosis.

Changes in the typology of Usher are related to genetic research. Gene identification has clearly shown that a number of different genes are involved in a wider range of 'types' of Usher than previously known. The symptomatic identification of Usher into – profoundly deaf, lose vision in teens (1) , moderately deaf, lose vision in twenties (2) , and lose both vision and hearing in thirties (3) does not match the genetic markers of different types. The new diagnoses shows that young people can be diagnosed with type 3 Usher and lose vision very quickly. People who thought they had Usher 1 may have Usher 2. One person who took part in an interview was diagnosed with Usher 2 (by genetic diagnosis) but did not have hearing loss. The 'prognoses' associated with the typology are no longer helpful, when they do not match the symptoms.

Obviously genetic research is important in the search for treatments and therapies for Usher and for RP. For this, people will need to know a

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medical 'type' of Usher, linked to the genes which are causing the problems. But for education, for independence and for planning, the symptomatic typologies may be the closest match to what actually *happens* for people with Usher. It is beyond the scope of this study to suggest how this should be resolved, but it may be a step in the right direction if the simple descriptors are used less frequently and with less emphasis.

Changes in education are linked probably most strongly to cochlear implant technology, but also to a more inclusive educational environment. 12 adults with Usher 1 all attended special schools for deaf pupils – only a quarter of them signing schools, though all now use BSL or SSE as their main communication. Although the numbers were small, of the 5 young people, 4 had cochlear implants, and were English language users – two attended mainstream schools, and two specialist schools which included the use of BSL. A higher proportion of the young people with Usher 2 attended mainstream schools than the proportion of adults who had. This suggests that more pupils were included – partly because they could be – in more recent times.

While it is clear that we did not interview any young people who did not know they had Usher, and did not try to reach anyone who did not, it does appear that it might be the case that at least some young people know about having Usher when older people, at the same age, might not have known. There seemed to be more openness. Genetic testing and other tests not developed perhaps 50 years ago, meant that a diagnosis of Usher could be more certain at a younger age.

14.2 The impact of diagnosis

One of the key aims of the project was to look at the impact of diagnosis on people immediately and over time. As has been found in many other studies, participants' perceptions of doctors breaking bad news was not good (e.g Right from the Start – Leonard, 1999). The impact of the diagnosis was for some the start of depression and struggle. But for others it was a spur to do more, and to others it was something to put aside for now.

Some people living with Usher, treated it as a thing – external to themselves, for others, as a way of being – integral to who they are. For those who treated it as a thing, Usher was something in their lives, something which they carried with them all the time, and to that extent it was part of them. However it did not define who they were, or what they did. They might have to get round it, avoid it, or sometimes face it head on and accept its presence, but they did not take it into themselves.

For others, it was a way of being. Usher was part of them, it was one of the things which identified them, which they saw as key to who they were. They might of course have difficulties with this, but it was still one of the things that made them themselves. Some of them, like Glen (in Schneider, 2006) embraced it and would not have chosen to be different. Their life would not be exactly the way they had envisaged it but that did not mean it had to be less.

Participants often embodied both of these positions simultaneously. Sometimes Usher was a thing – an eye condition, a hearing impairment, something which they could not do anything about, which they chose to ignore, and at other times sought to challenge. Yet at the same time Usher was central to their identity and made them who they were.

The impact of Usher as a disability – the point at which visual impairment became a problem – is sometimes also the point at which deafness came to mean something. They had not felt deaf, they had managed deafness, until they also could not see very well. Although to others their deafness was the same, to them, it had changed. For Deaf people, who used BSL, at the point at which they started to lose their vision, they began to feel less a part of the Deaf community and less in touch with BSL. They had embraced Deafness as part of them, but they felt it now as something they were less part of. Kyle and Barnett, (2007) show that deafblind people, including people with Usher, felt very excluded from Deaf communities. Hartmann, Sturley and others echo this in their blogs.

Visual impairment exacerbates hearing impairment. Mobility for a visually impaired person presents challenges, but as a dual sensory impaired person it presents far more. When swimming, people had to take off their hearing aids- and now that they could no longer see well,

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they could no longer manage the information around them. Communication became difficult when they could no longer lip read easily. People with Usher may not have felt deaf before, but now they were **deafblind**. Support was related to gadgets which helped hearing impaired people or visually impaired people, or strategies which helped one or the other –there was little which embraced deafblindness.

14.3 The Usherness of Usher

Throughout the text we have been looking at three features of Usher syndrome which have been key to our understanding of how it affects people's lives. These are; Change, Predictability/Uncertainty and Diversity.

We will now look at these three topics as they present across all the areas as a summary.

14.3.1 Change

Change is at the heart of Usher; but it is also at the heart of life. Hartmann (no date) talking about her adaptation (and her partners) to Usher says;

Change is a constant of life. If we don't change, we die.

There were changes throughout people's lives and some took on board the changes of Usher in much the same way as they took on the changes brought about by getting a job, having a child, or becoming ill with some other condition. They gathered up and went on. But there were certain aspects of change which were crucial to the experience of people with Usher. For some, change came at the moment of diagnosis, but not in the sense that anything changed except themselves, their self perception and their security. For most people things changed in relation to Usher when something happened which they could not avoid – when they stopped driving or started to read braille. These moments, rather than the moment of diagnosis, were ones when they needed support, which was often available only through family and friends. There were a number of occasions when people were forced to change not so much because of themselves but because of the inability of the

environment to adapt. People had to change schools, sometimes they were pleased and sometimes not, when the school they were at could no longer adapt to their levels of vision and hearing loss. Some people had to change jobs or leave work when workplace environments and managers were no longer prepared to change to meet their needs. People's lives changed when they gave up, or took up certain activities which they enjoyed, because of failing vision.

Their lives changed in small ways too, turning on the lights (with bigger electricity bills) or running hand in hand (instead of alone). Smaller changes maybe, but still very significant to the people who were making these changes because of the unpredictability associated with Usher. Or they might be positive changes; finding friends in support groups or deciding to have a baby or go quad biking **now**.

Interestingly, one of the things which did not change much was their communication methods. Of those who had Usher 1 but had limited access to BSL at school, all had gone on to use BSL as their preferred communication. These were however older adults, and it could be argued that this is unlikely to happen now; either they might have cochlear implants or they might go to a bilingual school. Those who were spoken English users stayed with spoken English, using hearing aids, implants or other means of amplification. They used some signs in the swimming pool or deafblind manual in cinemas, but they were not considering learning BSL, for the most part. Those who were BSL users might be considering using hands on sign – they might already use this in some situations. Some had to make changes related to very poor sight – one young person was using braille for reading and two older people were using it, or BSL users using hands on methods almost entirely.

They also all **knew** they would change – and dealt with this in the diverse ways that people with Usher proceeded with their lives. Some might decide to get on with it, and cope with each change, through new aids, new strategies, new support. Others might hang on as long as possible in the old way before an accident or similar forced them into something new.

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The constant and largely unpredictable change did place a strain on people's lives, with some being depressed for a time, and others being frustrated at new difficulties. This challenged their ontological security. But it was understood by the participants, in general, to be part of Usher – that they had to be ready to adapt – 'up and at 'em' (as Kat's mother said).

14.3.2 **Predictability/Uncertainty**

Hartmann (no date) writes of her change of relationship

Of course, not everyone knows what they're in for. It's a blessing and a curse to know. It's a blessing because it forces you to cherish what you have earlier in your life; a curse since you can't enjoy your sense as freely as others because you're constantly monitoring your losses.

Uncertainty is a theme underlying all the participants' experiences of Usher. Predictions had been made, frequently, especially at diagnosis, but in many cases, they had turned out to be inaccurate, or at least inaccurately phrased. Predictions based on the type of Usher were also inaccurate, based on the previous typologies.

There are particular issues which people could predict were likely to happen, but they could not tell when. These included, giving up driving (if they had ever started), using a cane, and giving up (if they had ever done them) certain ball sports. But exactly when these things would happen was not predictable for any individual, nor was it based on Usher type, nor on others in the family. Worrying about what would happen at these stages was noted by most of the participants at some time – even if their response was to shrug it off.

Others decided to beat the predictions, by starting in advance – by deciding to have a family quickly, to go travelling, to jump out of an aeroplane (attached to a parachute). In this way, they were challenging the predictions made and the fact that they might not be able to do things.

Uncertainty about what would happen was certainly a feature of living with Usher. The lack of consistency and the difficulty with establishing

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routines, because of constant changes, did have an effect on how people lived their lives. Interestingly, some did not want this to mean that they simply became more dependent – they did not want to become more patient or more sensitive. They wanted to take control, others looked into the future and resigned themselves to it, finding that they managed the unpredictability better if they rolled with the punches.

As difficult as the uncertainty of what would happen to them, is the uncertainty of how they would deal with it. They did not know what they would think, or how they would manage. But most of them did manage, and did remain thinking positively.

Also unpredictable was how others would behave. Colleagues, family and friends had to change as well, - they thought partners might leave, or colleagues might reject them (leading to some hiding Usher from work companions) or that friends would not want to be support workers as well as friends. But in fact, in many cases, they did continue to have friends, their partners did not leave them, though colleagues in mainstream working environments did not seem, overall, to be very helpful.

14.3.3 **Diversity**

Finally, the participants and their experiences are and continue to be diverse. Of course their lives are diverse; they are men and women, young people, adults and older adults, they come from all over the country and from a range of ethnic groups and backgrounds They are working, unemployed and studying, they are married and single, have children or don't. They are happy, sad, optimistic, worried, have good self image and esteem, weaker self esteem or a typical amount. It would be more surprising if their lives were all very similar.

Some of the experiences are quite negative and people differ in how depressed they were, how isolated and the extent to which they have given up activities. They differ in the amount of support they got (despite legislation to suggest otherwise). They had different experiences and some were told about Usher in very unhelpful ways. Some were finding adapting hard, and had not, for instance, been able to find work, or find

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leisure activities, or found only very limited access to the outside world. Some were bullied at school or excluded.

But in other ways the experiences are positive. There are many things which suggest that people with Usher can do many of the things they want to – they sometimes have to find a way round it – but they can. They manage change and so take part in blind cricket, managing their own businesses, attending mother and toddler groups, undertaking adventurous sports and having successful relationships.

The progress of their visual impairment (and hearing impairment) is also very diverse, and not linked to the predictors of type or other family members' experiences. Some are managing with very little sight at as young an age as 15, while well on in their 50s, others are still using sight to get around. Individual diversity is important in managing their progressive impairment, - what was an obstacle for one person might not be for another. Levels of vision and hearing loss were not good predictors for many of the concerns – work, school, families, activities; apart from obvious differences (such as taking part in sports for the blind rather than mainstream sports) the individual's own response was a much more important factor.

If asked about Usher, people would say different things. Their experiences show they felt about Usher in different ways. They would choose different support and different timings of support. They would give different advice to other people and they would expect different things.

14.4 What people with Usher may need

Through the research, a number of issues arose which indicated things that people with Usher might need. Of course, as throughout, diversity is key to this process. Not all people with Usher wanted the same things. What suits someone who has embraced Usher may not suit someone who is trying to live as if it isn't going to happen.

However, Usher does have an impact on school, on work, on family, and more.

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At the point at which they are diagnosed, and shortly after, the stories of our participants suggested that they

- wanted to know that there are specialists. They wanted the reassurance of someone who understood Usher and who could tell them what the latest developments were. They wanted to know what Usher was, and to know that someone else knew more than they did
- wanted to know what 'going blind' means. They may have been told they would 'go blind' but not understand that in fact most people with Usher do not lose all their vision very quickly, if indeed at all. The fact they had Usher would not inevitably mean they would have to learn braille (only three of our participants, ranging from 14-59 used braille, and one of these was a young person) although it would be likely to mean that at some point they would have to consider using a cane. Specialists cannot tell any individual what exactly will happen to them, but they can give some general advice or statistics which indicate that Usher is not necessarily what people might imagine by 'going blind'.
- wanted a level of support appropriate to their level of vision *now*. They did not need to learn braille when they were diagnosed, but they needed to understand some things about visual impairment. They needed information at the point of diagnosis, but they might need support later – counselling, advice, and information about resources they could use. Because Usher, unlike a medically managed illness, does not require frequent returns to doctors, they might not get in touch with anyone who can support them again for a long time. When and if they need to learn braille, they need to know how to get that help.

They needed support for their daily lives. In many cases, people had to find their own resources and their own support networks. Sense was a clear central provider of support for some, while others had little contact.

- Some participants did not want to be signed off as 'sick'. They were not being treated and the experience of others with Usher suggests that with appropriate adaptations, they could continue to work. Advice on Access to Work and employment legislation and

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support is probably a more useful response. Accompanying this, participants wanted better careers advice at school, especially if they needed to retrain.

- Some participants found organisations such as the ‘Young Professionals’ group from ‘RP Fighting Blindness’ to be very useful. Specific groups of peers related to their own age and their own level of visual difficulty were definitely the option preferred by some. Networks of informal support, whether meeting in person or online were as helpful for some as support from formal agencies set up to do this.
- In schools participants wanted access to support and advice from teachers specially trained in deafblindness who knew about the varied needs of people with Usher and who could tailor this to individual requirements.
- Some participants wanted detailed information on the genetic inheritance of Usher, this was particularly so for those who were planning a family. Readily available genetic tests to determine type and subtype were requested by some – specifically those who wanted to take part in gene therapies, or were finding it difficult to deal with the uncertainty of not having a clinical diagnosis.
- Participants also identified that partners, parents, friends, and siblings, were also in need of support and advice from specialist services and networks who understood the Usherness of Usher.
- Greater awareness of hidden disabilities, including communication and mobility strategies to support people with hearing and visual impairments were recommended. This was viewed as particularly important for hospital and medical staff, benefits agencies, public transport operatives, as well as the general public.
- Most significantly, overall was a desire by participants to share with other people with Usher, especially the newly diagnosed, their success, achievements, strategies for coping, and the knowledge that although it is tough, everything will be ok in the end.

14.5 What Next?

This study is a significant piece of research that explores the lives and experiences of people with Usher syndrome. This report and its findings – what it is like to live as a person with Usher in the UK today, the impact of diagnosis, and the Usherness of Usher (change, predictability/uncertainty, diversity) – lay the foundations for further rigorous, academic research on Usher. Subsequent research may include:

- An expansion of this original project to include people with Usher in their 70s, 80s and beyond. Also with careful attention paid to ethical issues, young children could be successfully involved in a similar study.
- Action research, locating people with Usher more firmly in the research process.
- As advances are made in relation to gene therapy and retinal implants an exploration of issues that arise from this.
- In-depth analysis of communities where there are higher incidences of Usher e.g. families
- Collaborative research with international colleagues

14.6 Final Comments

Bringing this report to an end, through it, we have presented the lives of people with Usher syndrome, showing the impact of the diagnosis on their experiences; education, communication, employment, friends and family, mobility – across all areas of their lives. There is no single message – the message belongs to each individual who took part in the project. To finish then we reflect on the nature of Life and Change with Usher in the words of two of the participants;

You just have to see what happens, when it happens, if it happens, but if you've got support, you can't ask for much more. (Chris, 33, type 3.)

I could worry about it on my own but I don't. You never know what's going to happen around the corner. Just get on with it. (Carolyn 39 type 2).

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